Sat. Jul 8, 2017

Poster Presentation Area

JCK Poster

JCK Poster 1 (II-JCKP1)

Basics/New Insights/Others

Chair:Tran Cong Bao Phung(Cardiology Department, Children Hospital 1, Ho Chi Minh City, VietNam)
6:15 PM - 7:15 PM Poster Presentation Area (Exhibition and Event Hall)

[II-JCKP1-01] Is younger the better for cardiac remodeling with transcatheter ASD closure in adult patients regardless of diastolic dysfunction?

○Lucy Youngmin Eun, Eui Young Choi, Young Won Yoon, Jae Young Choi （Department of Pediatric Cardiology, Yonsei University, Seoul, Korea）

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[II-JCKP1-02] Establish Heparin / Collagen-REDV selective active interface on ePTFE to promote endothelialization

○Yaping Shan, Bing Jia （Department of Children's Hospital ,FUDan University, Shanghai,China）

6:15 PM - 7:15 PM

[II-JCKP1-03] Vegfa signaling regulates diverse artery/vein formation in vertebrate vasculatures

○Diqi Zhu[1], Daqing Jin[2], Yabo Fang[2], Yiwei Chen[1], Weijun Pan[3], Dong Liu[4],Fen Li[1], Tao Zhong[2], （1.Department of Pediatric Cardiology , Shanghai Children's Medical Center Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai, China, 2.State Key Laboratory of Genetic Engineering, Zhong Shan Hospital, School of Life Sciences, Fudan University, Shanghai, China, 3.Institute of Health Sciences, Shanghai Institutes for Biological Sciences, Chinese Academy of Sciences, Shanghai, China, 4.Co-innovation Center of Neuroregeneration, Jiangsu Key Laboratory of Neuroregeneration, Nantong University, Nantong, China）

6:15 PM - 7:15 PM

[II-JCKP1-04] Efficacy of a Special 6-Dose Palivizumab Prophylaxis Protocol for Respiratory Syncytial Virus Infection in Congenital Heart Disease in Subtropical Areas

○Yu Chuan Hua[1], Shuenn Nan Chiu[2], Jou Kou Wang[2] （1.Cardiac Children's Foundation, Taipei, Taiwan, 2.Department of Pediatrics, National Taiwan University Hospital and Medical College, National Taiwan University, Taipei, Taiwan）

6:15 PM - 7:15 PM

[II-JCKP1-05] Assessment of Right Ventricular Systolic Function in Patients after Repair of Tetralogy of Fallot with Various Degrees of Pulmonary Regurgitation

○Manatomo Toyono[1], Shunsuke Yamada[1], Mieko Aoki-Okazaki[2], Tsutomu Takahashi[1] （1.Department of Pediatrics, Akita University, Akita, Japan, 2.Interconnected Medical Education and Support Systems, Akita University, Akita, Japan）

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[II-JCKP1-06] TGF-β/Smad3 signaling promotes collagen synthesis in PASMC through down-regulating miR-29b

○Ting Ting Xiao, Juan Chen （Department of Cardiology,Shanghai Children's Hosptial, Shanghai, China）

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[II-JCKP1-07] T-VDCC CaV3.1 and CaV3.2 involves in pulmonary hypertension

○Jie Shen[1], Zixu Huang[2], Yuanyang Wang[2] （1.Department of Cardiology, Shanghai Children's Medical Center affiliated to Shanghai Jiaotong University School of Medicine, China, 2.Department of Cardiology, Shanghai Children's Hospital, Shanghai Jiaotong University, China）

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[II-JCKP1-08] Pleural effusion as complication of middle aortic syndrome treated by stent implantation

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[II-JCKP1-09] Effects of respiratory ciliary dysfunction on increased postoperative respiratory complications in congenital heart disease patients with heterotaxy

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[II-JCKP1-10] Successful double intraventricular reroutings and Dams Kaye Stansel anastomosis as Rastelli modification for rare formed double outlet right ventricle

○Takaya Hoashi[1], Isao Shiraishi[2], Kenichi Kurosaki[2], Masatoshi Shimada[1], 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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JCK Poster

JCK Poster 2 (II-JCKP2)
Kawasaki Disease/General Cardiology/Adult Congenital Heart Disease

Chair:Kaiyu Zhou(Department of Pediatric Cardiology, West China Second University Hospital, Sichuan University, Chengdu, Sichuan, China)

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[II-JCKP2-01] Progressive Coronary Dilatation for Eighteen Months in Kawasaki Disease: A Case Report

○CHIAHUI WU, Ming-Tai Lin, Jou-Kou Wang, Mei-Hwan Wu （Department of Pediatrics, National Taiwan University Hospital, Taipei, Taiwan）

6:15 PM - 7:15 PM

[II-JCKP2-02] Shock as prominent early manifestation of kawasaki disease in Children

○Kaiyu Zhou, Wang Chuan, Yimin Hua, Yifei Li （Department of Pediatric Cardiology, West China Second University Hospital, Sichuan University, Chengdu, Sichuan, China）

6:15 PM - 7:15 PM

[II-JCKP2-03] The level of serum TNF-a in intravenous immunoglobulin non-responsive children with Kawasaki disease

○Wang Yun[1], Cui Dai[2] （1.Department of pediatrics, Beijing new century women's and children's hospital, Beijing, China, 2.Children's Hospital Affiliated to Capital Institute of Pediatrics, Beijing, China）

6:15 PM - 7:15 PM

[II-JCKP2-04] Clinical features of Kawasaki Disease children with urinary tract involvement

○Wang Yang[1], Wang Yun[2], Zhou Nan[3], Chen Jia Jia[3], Chen Li[3], Tang Xiao Lei[3] （1.Department of Pediatrics,Beijing New Centery International Children's Hospital, Beijing, China, 2.Beijing New Century Women's and Children's Hospital, China, 3.Beijing Children's Hospital, China）

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[II-JCKP2-05] The increased serum levels of Interleukin-21 in Kawasaki disease

○Hae Yong Lee （Department of Pediatric Cardiology, Yonsei University,Wonju, Korea）

6:15 PM - 7:15 PM

[II-JCKP2-06] The relationship between children metabolic syndrome and target organ damage of essential hypertension

○Yao Lin, Lin Shi, Jie Mi, Yang Liu （Department of Cardiology, Capital Institute of Pediatrics, Beijing, China）

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[II-JCKP2-07] The effect of mother's social support on children with congenital heart disease and mother-child interaction on children's problem behavior.

○Ju Ryoung Moon[1,4,5,6], Jinyoung Song[1,2,4,5,6,7], June Huh[1,2,4,5,6,7], I-Seok Kang[1,2,4,5,6,7], Ji-Hyuk Yang[1,3,4,5,6,7], Tae-Gook Jun[1,3,4,5,6,7] （1.Grown-Up Congenital Heart Clinic, 2.Department of Pediatrics, 3.Department of Thoracic &Cardiovascular Surgery, 4.Cardiac Center, 5.Heart Vascular and Stroke Institute, 6.Samsung Medical Center, 7.Sungkyunkwan University School of Medicine）

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[II-JCKP2-08] Changes of content of circulating

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endothelial cells and endothelial progenitor cells in patients with primary hypertension in peripheral blood

○Lin Shi, Yuanyuan Zhang, Yao Lin, Yang Liu （Department of Cardiology,Capital Institute of Pediatrics, Beijing, China）

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[II-JCKP2-09] Role of Cardiopulmonary Exercise Testing for Prediction of Pregnancy Outcome in Women with Congenital Heart Disease

○Midori Fukuyama, Hideo Ohuchi, Chizuko Kamiya, Mitsuhiro Fujino, Kenichi Kurosaki, Jun Yoshimatsu （National Cerebral and Cardiovascular Center, Japan）

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[II-JCKP2-10] Myocardial Perfusion Abnormality by SPECT Imaging Correlates with Systemic Ventricular Property in Adults with Congenital Heart Disease

○Mitsuhiro Fujino, Hideo Ouch, Yosuke Hayama, Tohru Iwata, Jun Negishi, Aya Miyazaki, Etsuko Tsuda, Kenichi Kurosaki, （Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan）

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○Lucy Youngmin Eun, Eui Young Choi, Young Won Yoon, Jae Young Choi （Department of Pediatric Cardiology, Yonsei University, Seoul, Korea）

Background: With the transcatheter closure of atrial septal defect (ASD), it is noticeable for the change of left atrium and left ventricle at longterm follow-up. The purpose of this study was to assess the cardiac remodeling with the aspect of diastolic function in adult ASD patients.
Methods and results: Asymptomatic thirty eight patients(48.6±17.1 years) on the diagnosis of ASD from health check-up who underwent ASD device closure were enrolled. The defect size was 21.77+6.79 mm. The medical records were reviewed and divided into two groups : I <50year old (33.06±9.43), II≥ 50 year old(62.55±7.54). The echocardiographic data in between pre-closure and follow-up showed significant differences at LVEDD(40.76±3.28 vs 43.39±3.52, p<0.001), LV mass(99.64±28.81 vs 116.57±32.03, p <0.01), and RV pressure(36.88±12.20 vs 31.81±11.11, p=0.04). Then, at post-closure follow-up, tissue Doppler measurements were significantly decreased and E/E'(8.79+3.19 vs 11.58±4.80, p<0.005) was more elevated than pre-closure. Between the two groups, mitral A, tissue Doppler E', A', S' were all decreased, and E/E'(pre : 7.41±1.42 vs 9.60±5.15, p<0.003, post : 10.49±3.95 vs 13.03+4.05, p<0.02) was much higher at group II at both pre-closure and longterm follow up as well.
Conclusion: After the transcatheter ASD closure in adults, it might be masked for severe diastolic deterioration. Relative younger age might be better for the remodeling to protect myocardial function after transcatheter closure of ASD. Thus, it may benefit to close ASD as earlier age as possible in adults.

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[II-JCKP1-02] Establish Heparin / Collagen-REDV selective active interface on ePTFE to promote endothelialization

○Yaping Shan, Bing Jia （Department of Children's Hospital ,FUDan University, Shanghai,China）

Objective To construct heparin / collagen-REDV selective active interface on the surface of expanded polytetrafluoroethylene, and observe the endothelialization level and cell activity on the surface of ePTFE in vitro. **Methods** Five layers of heparin and collagen combined coating [$(HEP/COL)_5$] were prepared on the surface of 0.1mm ePTFE membrane by layer by layer self-assembly technique, and REDV peptides were coated on its surface ,then a selective active interface on ePTFE was obtained. Unmodified ePTFE、$(HEP / COL)_5$ modified ePTFE、$(HEP/COL)_5$-REDV modified ePTFE and $(HEP/COL)_5$-REVD modified ePTFE were co-cultured with umbilical vein endothelial cells for 24 hours to 72 hours , observed the endothelialization level and cell activity on ePTFE. **Results** Co-culture 1h and 6h, the number of endothelial cells adherent on $(HEP/COL)_5$-REDV modified ePTFE was more than other groups(P<0.05); Co-culture 24h、48h、72h,the endothelial cells on modified ePTFE were significantly more than unmodified , the endothelial cell number and cell activity was highest in $(HEP/COL)_5$-REDV modified ePTFE group(P<0.05). **Conclusions** Using layers by layer self-assembly technique to build a selective active interface on ePTFE surface can promote endothelial cells adherent to ePTFE , as well as

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promote cell proliferation and improve cell activity.

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[II-JCKP1-03] Vegfa signaling regulates diverse artery/vein formation in vertebrate vasculatures

○Diqi Zhu[1], Daqing Jin[2], Yabo Fang[2], Yiwei Chen[1], Weijun Pan[3], Dong Liu[4],Fen Li[1], Tao Zhong[2], （1.Department of Pediatric Cardiology , Shanghai Children's Medical Center Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai, China, 2.State Key Laboratory of Genetic Engineering, Zhong Shan Hospital, School of Life Sciences, Fudan University, Shanghai, China, 3.Institute of Health Sciences, Shanghai Institutes for Biological Sciences, Chinese Academy of Sciences, Shanghai, China, 4.Co-innovation Center of Neuroregeneration, Jiangsu Key Laboratory of Neuroregeneration, Nantong University, Nantong, China）

Objective— Vascular endothelial growth factor A (Vegfa) signaling regulates vascular development during embryogenesis and organ formation. However, the signaling mechanisms that govern formation of arteries and veins in various tissues are incompletely understood. We aimed to understand differential functions and mechanisms of Vegfa signaling during formation of diverse arteries/veins in zebrafish trunk and head vasculatures.

Approach and Results— We utilized TALEN to generate zebrafish *vegfaa* mutants. *vegfaa*$^{-/-}$ embryos are embryonic lethal, and display a complete loss of the dorsal aorta and expansion of the cardinal vein. We find that activation of Vegfa signaling expands the arterial cell population at expense of venous cells during vasculogenesis of axial vessels in the trunk. Vegfa signaling regulates endothelial cell proliferation after arterial-venous specification. Formation and extension of tip cell filopodia of intersegmental vessels are significantly inhibited in both Vegfa-deficient and Vegfa-overexpressing embryos.

Conclusions— Our results indicate that Vegfa signaling induces formation of the dorsal aorta at expense of the cardinal vein during vasculogenesis, and is required for angiogenic formation of mesencephalic veins and central arteries in the brain. These findings suggest that Vegfa signaling governs formation of diverse arteries/veins by distinct cellular mechanisms in vertebrate vasculatures.

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

[II-JCKP1-04] Efficacy of a Special 6-Dose Palivizumab Prophylaxis Protocol for Respiratory Syncytial Virus Infection in Congenital Heart Disease in Subtropical Areas

○Yu Chuan Hua[1], Shuenn Nan Chiu[2], Jou Kou Wang[2] （1.Cardiac Children's Foundation, Taipei, Taiwan, 2.Department of Pediatrics, National Taiwan University Hospital and Medical College, National Taiwan University, Taipei, Taiwan）

Objective: Palivizumab prophylaxis for respiratory syncytial virus (RSV) infection in patients with hemodynamically significant congenital heart disease (hsCHD) has been proven to be effective in RSV

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seasons in Western countries. To study the efficacy of a novel palivizumab prophylaxis protocol for hsCHD in subtropical areas without clear RSV seasonality.
Method: The Taiwan National Health Insurance has provided reimbursement for palivizumab prophylaxis with a special 6-dose monthly protocol for patients with hsCHD. We performed a study to assess the trend of RSV infection in patients .
Results: Totally, 1646 patients were enrolled: 763 in the study group and 883 in the control group. Cyanotic CHD was observed in 41.6% of the patients. After a mean of 3.9 doses of palivizumab injection, the RSV hospitalization rates and hospitalization days decreased by 50% and 54%, respectively, in the study group compared with the control group ($P = .013$ and .025, respectively), and these values decreased further by 68.7% and 70% ($P < .001$ and .003), respectively. The RSV hospitalization rates decreased from 4.4% in the pre-palivizumab period to 2% in the palivizumab period ($P = .038$). The efficacy of this protocol was prominent both in patients with cyanotic hsCHD and in those with acyanotic hsCHD. The main causes of RSV infection after the palivizumab period were noncompliance and a delayed diagnosis of CHD.
Conclusions: Palivizumab prophylaxis through the novel 6-dose monthly protocol for patients with hsCHD is effective in reducing RSV-related hospitalization, with less doses and cost.

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[II-JCKP1-05] Assessment of Right Ventricular Systolic Function in Patients after Repair of Tetralogy of Fallot with Various Degrees of Pulmonary Regurgitation

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Objective: The objective of the study is to compare advantage between tricuspid annular isovolumic acceleration (IVA) and tricuspid annular plane systolic excursion (TAPSE) for stratification of impact of pulmonary regurgitation (PR) on right ventricular (RV) systolic function in patients after repair of tetralogy of Fallot (TOF).
Methods: IVA was measured by dividing the myocardial velocity during isovolumic contraction by its acceleration time. TAPSE was measured using M-mode echocardiography through the lateral annulus of tricuspid valve in the apical 4-chamber plane. PR degree was assessed by the number of correspondence to the following conditions; 1) diastolic flow reversal in the main pulmonary artery, 2) diastolic flow reversal in the branch pulmonary arteries, 3) pressure half-time of PR signal <100 msec and 4) duration of the PR signal/total duration of diastole ratio <0.77. PR degree was graded from 0 to 4.
Results: Twenty-two patients were enrolled to the study. Age, female, period after the TOF repair and body height of the patients were 11 ± 6 years, 55%, 8 ± 5 years and 137 ± 28 cm, respectively. In all the patients, IVA and TAPSE were 160 ± 27 cm/sec^2 and 12 ± 2 mm, respectively. PR degree was graded as 2, 3 and 4 in 4, 14 and 4 patients, respectively. By the Kruskal-Wallis test, only IVA showed a significant difference among the 3 PR degrees.
Conclusion: IVA can be a useful index for the stratification of RV function in patients after TOF repair with various degrees of PR.

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[II-JCKP1-06] TGF-β/Smad3 signaling promotes collagen synthesis in PASMC through down-regulating miR-29b

○Ting Ting Xiao, Juan Chen （Department of Cardiology,Shanghai Children's Hosptial, Shanghai, China）

miR-29 is reported to be associated with fibrosis diseases in which collagen synthesis plays an important role and participates in the development of liver fibrosis, renal fibrosis, pulmonary fibrosis, cardiac fibrosis. Deposition of extracellular matrix (ECM), such as fibronectin and collagen, and proliferation, migration, and hypertrophy of vascular smooth muscle cells (VSMCs) result in PA hypertrophy and muscularization, leading to increased pulmonary vascular resistance in PAH. With the possible links among HPH, collagen, in this study we examined the role and therapeutic potential of miR-29 in rats model of pulmonary hypertension induced by MCT. In conclusion, miR-29b plays an important role in collagen synthesis and may be a therapeutic agent for PAH under the regulating of the TGF-β/Smad3 pathway.

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[II-JCKP1-07] T-VDCC CaV3.1 and CaV3.2 involves in pulmonary hypertension

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Pulmonary hypertension (PH) is associated with hyperreactivity to vasoconstrictor agents and remodelling of pulmonary arteries with proliferation and migration of pulmonary arterial smooth muscle cells (PASMCs). Intracellular Ca^{2+} regulates many cellular processes, such as cell cycle progression, proliferation and apoptosis. Voltage-dependent Ca^{2+} channels(VDCC) can regulate intracellular Ca^{2+} levels. L-, T- and P/Q-type channels have been identified in vascular smooth muscle cells. L-VDCC inhibitors are not so efficient in the treatment of PH. T-type channels have been cloned, and systematically named CaV3.1, CaV3.2 and CaV3.3 T-type channels, respectively. T-type calcium channel antagonists, mibefradil and NNC-55-0396 inhibit cell proliferation in leukemia cell lines. Chronic hypoxia selectively enhances T-VDCC activity in pulmonary artery. We hypothesize that T-VDCC could constitute an alternative therapeutic target in PH.
In our research, we find that the expression of CaV3.1 and CaV3.2 are up-regulated in MCT- or hypoxia-induced PAH. Inhibition of T-VDCC CaV3.1 and CaV3.2 suppresses the proliferation of PASMC during hypoxia by delaying the G1/S phase conversion, and inhibition of CaV3.1 and CaV3.2 alleviates progression of MCT-induced PAH in rats. Further research indicates that blockade of CaV3.1 and CaV3.2 may delay G1/S phase through p-ERK/CCND1 signaling pathway. These observations may provide new mechanistic insights into pulmonary hypertension.

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[II-JCKP1-08] Pleural effusion as complication of middle aortic syndrome treated by stent implantation

○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)

Objectives: describe a rare complication of stent implantation in one patient with long abdominal and thoracic aortic stenosis due to middle aortic syndrome
Methods: one case report
Result: Patient: 12 year old girl with upper limb hypertension due to long segment abdominal and thoracic aortic stenosis due to middle aortic syndrome. Intervention: Aortic radiography shows a 56mm long abdominal and thoracic aorta stenosis (about 6 cm below the aortic isthmus and 3 cm above the renal artery origin). Covered stent 61 x 12 was implanted at the lower part of the narrow segment. Aneurysm at the upper ending of stent was discovered after that. Another stent 61 x 12 was implanted to cover that aneurysm. However, a fistula between the two stent still exists. Balloon dilation at the junction site was performed. The fistula finally disappeared. There was no pressure gradient through the stent
Complication: massive right pleural effusion.
Cause:the aneurysm after first stent implantation drained into the right pleural cavity.
Conclusion:
Inflammation in Takayasu disease, long and severve stenosis are high risk factors for ruptures during stenting procedures. We suggest staged dilation by using long covered stents for those cases to prevent complications.

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[II-JCKP1-09] Effects of respiratory ciliary dysfunction on increased postoperative respiratory complications in congenital heart disease patients with heterotaxy

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

Ciliary motion play a critical role on both airway mucus clearance system and left-right body axis development.We investigated whether CHD-heterotaxy patients may have ciliary dysfunction(CD) and its effect on worse postsurgical outcomes.
We assessed 87 CHD patients with heterotaxy for airway CD, and 100 healthy persons were also recruited as controls. Videomicrocopy was used to examine ciliary motion in nasal tissue, and nasal nitric oxide (nNO) was measured; nNO level is typically low with Primary Ciliary Dysfunction(PCD).
40 patients (46%) exhibited CD characterized by abnormal ciliary motion among total 87 heterotaxy patients, compared with 1subject with CD among 100 health controls(1%). Among 40 heterotaxy patients with CD, 27 patients appeared below or near the PCD cutoff values, compared with all normal nNO levels in health controls. We examined postsurgical outcome in 40 heterotaxy-CHD patients with CD, compared with 32 heterotaxy-CHD patients without CD. We found mean length of postoperative

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hospital stay (14.1 vs 11.2 days; OR, 2.4) and mechanical ventilation(65 vs 53 hours; OR, 2.1) were significantly increased in the heterotaxy patients with CD. Also elevated were number of reintubation(1.8 vs 1.3; OR, 3.1), salvage (11.5% vs 5.1%: OR, 2.1).
Our studies show that CHD patients with heterotaxy have substantial risk for CD and increased respiratory disease. Heterotaxy-CHD patients with CD may have increased risks for respiratory deficiencies. Overall, there was a trend toward increased mortality in CD patients with intermediate follow-up evaluation.

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[II-JCKP1-10] Successful double intraventricular reroutings and Dams Kaye Stansel anastomosis as Rastelli modification for rare formed double outlet right ventricle

○Takaya Hoashi[1], Isao Shiraishi[2], Kenichi Kurosaki[2], Masatoshi Shimada[1], 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）

Fetus echocardiography detected her double outlet right ventricle (DORV) with subpulmonary ventricular septal defect (VSD) at the gestational age of 27 weeks. She was born at 38 weeks of gestational age by normal vaginal delivery, and the echocardiography diagnosed doubly committed VSD without pulmonary stenosis, however, multi-slice computed tomography (MSCT) suspected VSD was located at subaortic to subpulmonary area widely, but which was seemed to be divided by conus septum. Blloon atrial septectomy was preceded at the age of 11 days, then which was followed by pulmonary artery banding at the age of 15 days. MSCT was again performed at the age of 12 months, and 3D heart replica was made for surgical simulation. Aorta was located at posterior right side of pulmonary artery, so VSD must be located at subaortic area, but which was restrictive. Interestingly, large subpulmonary VSD was additionally existed and 2 VSDs were clearly divided by well-developed conus septum. After confirmation of sufficient size of both ventricular cavity at the age of 15 moths by cardiac cineangiography, intra-ventricular rerouting (IVR)s from subpulmonary VSD to native pulmonary valve and subaortic VSD to native aortic valve with expanded polytetrafluoroethylene patches, Damus-Kaye Stansel anastomosis, and placement of right ventricle to pulmonary artery conduit with 14mm Contegra® was performed as the complete biventricular repair. Postoperative course was uneventful, and echocardiography showed both ventricular functions were well preserved without both outflow tract obstructions.

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JCK Poster

JCK Poster 2 (II-JCKP2)

Kawasaki Disease/General Cardiology/Adult Congenital Heart Disease

Chair:Kaiyu Zhou(Department of Pediatric Cardiology, West China Second University Hospital, Sichuan University, Chengdu, Sichuan, China)

Sat. Jul 8, 2017 6:15 PM - 7:15 PM Poster Presentation Area (Exhibition and Event Hall)

[II-JCKP2-01] Progressive Coronary Dilatation for Eighteen Months in Kawasaki Disease: A Case Report

○CHIAHUI WU, Ming-Tai Lin, Jou-Kou Wang, Mei-Hwan Wu （Department of Pediatrics, National Taiwan University Hospital, Taipei, Taiwan）

6:15 PM - 7:15 PM

[II-JCKP2-02] Shock as prominent early manifestation of kawasaki disease in Children

○Kaiyu Zhou, Wang Chuan, Yimin Hua, Yifei Li （Department of Pediatric Cardiology, West China Second University Hospital, Sichuan University, Chengdu, Sichuan, China）

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[II-JCKP2-03] The level of serum TNF-a in intravenous immunoglobulin non-responsive children with Kawasaki disease

○Wang Yun[1], Cui Dai[2] （1.Department of pediatrics, Beijing new century women's and children's hospital, Beijing, China, 2.Children’s Hospital Affiliated to Capital Institute of Pediatrics, Beijing, China）

6:15 PM - 7:15 PM

[II-JCKP2-04] Clinical features of Kawasaki Disease children with urinary tract involvement

○Wang Yang[1], Wang Yun[2], Zhou Nan[3], Chen Jia Jia[3], Chen Li[3], Tang Xiao Lei[3] （1.Department of Pediatrics,Beijing New Centery International Children's Hospital, Beijing, China, 2.Beijing New Century Women’s and Children’s Hospital, China, 3.Beijing Children’s Hospital, China）

6:15 PM - 7:15 PM

[II-JCKP2-05] The increased serum levels of Interleukin-21 in Kawasaki disease

○Hae Yong Lee （Department of Pediatric Cardiology, Yonsei University,Wonju, Korea）

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[II-JCKP2-06] The relationship between children metabolic syndrome and target organ damage of essential hypertension

○Yao Lin, Lin Shi, Jie Mi, Yang Liu （Department of Cardiology, Capital Institute of Pediatrics, Beijing, China）

6:15 PM - 7:15 PM

[II-JCKP2-07] The effect of mother's social support on children with congenital heart disease and mother-child interaction on children's problem behavior.

○Ju Ryoung Moon[1,4,5,6], Jinyoung Song[1,2,4,5,6,7], June Huh[1,2,4,5,6,7], I-Seok Kang[1,2,4,5,6,7], Ji-Hyuk Yang[1,3,4,5,6,7], Tae-Gook Jun[1,3,4,5,6,7] （1.Grown-Up Congenital Heart Clinic,

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2.Department of Pediatrics, 3.Department of Thoracic &Cardiovascular Surgery, 4.Cardiac Center, 5.Heart Vascular and Stroke Institute, 6.Samsung Medical Center, 7.Sungkyunkwan University School of Medicine）

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[II-JCKP2-08] Changes of content of circulating endothelial cells and endothelial progenitor cells in patients with primary hypertension in peripheral blood

○Lin Shi, Yuanyuan Zhang, Yao Lin, Yang Liu （Department of Cardiology,Capital Institute of Pediatrics, Beijing, China）

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[II-JCKP2-09] Role of Cardiopulmonary Exercise Testing for Prediction of Pregnancy Outcome in Women with Congenital Heart Disease

○Midori Fukuyama, Hideo Ohuchi, Chizuko Kamiya, Mitsuhiro Fujino, Kenichi Kurosaki, Jun Yoshimatsu （National Cerebral and Cardiovascular Center, Japan）

6:15 PM - 7:15 PM

[II-JCKP2-10] Myocardial Perfusion Abnormality by SPECT Imaging Correlates with Systemic Ventricular Property in Adults with Congenital Heart Disease

○Mitsuhiro Fujino, Hideo Ouch, Yosuke Hayama, Tohru Iwata, Jun Negishi, Aya Miyazaki, Etsuko Tsuda, Kenichi Kurosaki, （Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan）

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[II-JCKP2-01] Progressive Coronary Dilatation for Eighteen Months in Kawasaki Disease: A Case Report

○CHIAHUI WU, Ming-Tai Lin, Jou-Kou Wang, Mei-Hwan Wu （Department of Pediatrics, National Taiwan University Hospital, Taipei, Taiwan）

Background

Kawasaki Disease (KD) is one of the most common vasculitis in childhood and may lead to cardiac sequelae. Progressive coronary dilatation is not uncommon for KD patients with giant coronary aneurysms, but aneurysms rarely keeps enlarging one year after fever onset.

Case report

We, herein, report a 6-year-old KD girl who suffered from KD at her 4.5 years. She had immunoglobulin at 9 days after fever onset and fever subsided soon. The echocardiogram before IVIG demonstrated LMCA 2.9mm (Z-score +1.85), LAD 3.06mm (Z-score +2.18), proximal RCA giant aneurysm, 8.3mm (Z-score +6.94). Follow-up echocardiography showed her RCA gradually dilated from the initial 8.3mm to 11.2mm (4Y7M) and gradually to 16.1mm (6Y). She had aspirin, low molecular weight heparin, propranolol, atorvastatin (4Y7M-4Y9M) and captopril. At her age of six, cardiac perfusion scan didn't show any perfusion defect.

Review of Literature

We reviewed the literature and found another three similar cases. One was from Japan and his right coronary aneurysm enlarged gradually from 7.8mm to 19.5mm over the next 17 years. The third cases came from United States. She had a progressively enlarged LAD aneurysm that measured 7mm at the time of initial KD diagnosis and 25mm at the time of surgery 11 years later.

Summary

Giant coronary aneurysms with progressive coronary dilatation lasting longer than 1 year are rare. Optimal managements are still unclear in the unique subgroup of KD patients. Multi-center registry may clarify their real outcomes and suggest the future directions.

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[II-JCKP2-02] Shock as prominent early manifestation of kawasaki disease in Children

○Kaiyu Zhou, Wang Chuan, Yimin Hua, Yifei Li （Department of Pediatric Cardiology, West China Second University Hospital, Sichuan University, Chengdu, Sichuan, China）

OBJECTIVEWe sought to define the characteristics that distinguish Kawasaki diseaseshock syndrome (KDSS)from hemodynamically normal Kawasaki disease. **METHODS**We collected data prospectively for all patients with Kawasaki disease whowere treated at a single institution during a 2-year period. We compared clinical and laboratory features, coronary artery measurements,and responses to therapy and analyzed indices of ventricular systolic anddiastolic function during acute and convalescent Kawasaki disease.

RESULTS Of 231 consecutive patients with Kawasaki disease, 4 met the definitionfor KDSS. All required fluid resuscitation and vasoactive infusions. Compared with patients without shock, patientswith Kawasaki disease shock syndrome were more often female,the age of patientswith KDSS was between

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5.2 to 8.9 years, as well as had largerproportions of bands, higher C-reactive protein concentrations, and lower hemoglobinconcentrations and platelet counts. Evidence of consumptive coagulopathy wascommon in the KDSS group. Patients with KDSS more often had impaired left ventricular systolic function, mitralregurgitation , coronary arteryabnormalities, and intravenous immunoglobulin resistance . Impairment of ventricular relaxation and compliance persistedamong patients with KDSS after the resolution of other hemodynamic disturbances.
CONCLUSIONSPatients with KDSS may have uneven clinical course and may be misdiagnosed in thebeginning. They may have more prominent inflammatory markers in the early phase and higherrisk of coronary artery dilatation.

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[II-JCKP2-03] The level of serum TNF-a in intravenous immunoglobulin non-responsive children with Kawasaki disease

○Wang Yun[1], Cui Dai[2] (1.Department of pediatrics, Beijing new century women's and children's hospital, Beijing, China, 2.Children's Hospital Affiliated to Capital Institute of Pediatrics, Beijing, China)

Objective Study TNF-a levels in 131 KD patients, help to identify risk factor of IVIG non-responsiveness, and explore further treatment. **Methods** KD patients were divided into non-responsive and sensitive group, 28 healthy and 16 fe- brile patients were also recruited. **Results** 1) Proportion of CAA (30.0% vs. 7.2%) in the non-responsive group was significantly higher than those in sensitive group ($p<0.01$); Proportion of cardiomegaly in the non-responsive group was also significantly higher than that in the sensitive group at acute phase ($p<0.01$); IVIG non-responsiveness was an independent risk factor for cardiovascular complications.2) The high TNF-a levels existed in KD, and there was a statistical difference in KD patients compared with healthy children, and febrile controls, respectively ($p<0.01$). 3) Before therapy, TNF-a was 128.65 (97.45, 251.88) pg/ml VS 44.20 (21.00, 125.78) pg/ml; there was a statistical difference within non-responsive and sensitive group ($p<0.01$). The elevation of TNF-a continually existed in the non-responsive group after initial IVIG , there was also a statistical difference between them ($p<0.05$). 4) Male, TNF-a>100 pg/ml and ALB were independent risk factors associated with IVIG re-treatment. **Conclusions** High TNF-a levels existed in KD. IVIG could allow TNF-a levels reduce, but TNF-a were continually elevated in the non-responsive group after initial IVIG. IVIG non-responsiveness was an independent risk factor for cardiovascular complications. Risk factors associated with IVIG re-treatment include Male sex and the high level of TNF-a.

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[II-JCKP2-04] Clinical features of Kawasaki Disease children with urinary tract involvement

○Wang Yang[1], Wang Yun[2], Zhou Nan[3], Chen Jia Jia[3], Chen Li[3], Tang Xiao Lei[3] (1.Department of Pediatrics,Beijing New Centery International Children's Hospital, Beijing, China, 2.Beijing New Century Women's and Children's Hospital, China, 3.Beijing Children's Hospital, China)

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Objective:Retrospective797 KD children, try to explore the clinical characteristics of KD children with urinary tract involvement.
105 Patients with urinary tract involvement as the study group, 98 urinary tract non-involvement as control, analyzed the clinical features, inflammatory and sensitivity to IVIG, as well as in children with different degree of urine test, within these groups .
Results: 1) 13.2% (105/797) KD children were urinary tract involvement, gender ratio was 2:1. Abnormal urine test shows that 70.5% (74/105) of urinary tract involvement KD children had the white blood cells (several ~ full vision/HP). 67 cases underwent ultrasound and showed 3 cases with bilateral renal enlargement and 5 cases with echogenicity. 2) The CRP level is significantly higher in study group than the control group, but ESR has only showed a rising trend without significant difference. 16.2% patients was not sensitive to IVIG in Urinary tract involvement group, there was no significant difference between the two groups, although 7.2% higher than that of control group. 3) 20.6% patients in hematuria and/ orproteinuria group is not sensitive to IVIG, but there was no significant difference within groups.
Conclusion: KD patients with urinary tract involvement were more common in infants and young children, white blood cells was often seen in the urine. The inflammatory indexes increased in the patients with urinary tract involvement. The percent of non-sensitive to IVIG had the higher trend in the urinary tract involvement group, especially the patients with hematuria and proteinuria.

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[II-JCKP2-05] The increased serum levels of Interleukin-21 in Kawasaki disease

○Hae Yong Lee (Department of Pediatric Cardiology, Yonsei University,Wonju, Korea)

Purpose: It has been reported that serum level of immunoglobulin E (IgE) is increased in patients with Kawasaki disease (KD) after acute phase. We investigated whether the interleukin-21 (IL-21) could be related with the high IgE in KD.
Methods: From June 2008 to June 2010, 49 patients with KD admitted in Wonju Christian Hospital and 13 controls with high fever due to unknown infection who had no history of KD were included in this study. The sera from patients and controls were collected and checked in terms of immunoglobulin E (Chemiluminescent method, Siemens, Munich, Germany) and IL-21 (ELISA, eBioscience, San Diego, USA).
Results: The group of patients with KD was composed of 39 complete KD and 10 incomplete KD. Among patients with KD, 10 patients had coronary arterial dilatation (CAD) and 39 patients had no coronary complications. The median value of IL-21 in patients with KD was significantly increased as 466 pg/mL (range: 0-1544) while that value in controls was <62.5 pg/mL (range: 0- 825 pg/mL) (P <0.01). We could not find the significant correlation between the serum level of IgE and that of IL-21 in patients with KD (Spearman R=0.2, P = 0.08) though 30% of patients with KD showed increased IgE more than 100 IU/mL. In addition, our data showed no significant difference between CAD group and non CAD group in terms of serum IL-21.
Conclusion: Our data showed that IL-21 is increased in patients with KD. There was no significant correlation between high IgE and the level of IL-21.

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[II-JCKP2-06] The relationship between children metabolic syndrome and target organ damage of essential hypertension

○Yao Lin, Lin Shi, Jie Mi, Yang Liu （Department of Cardiology, Capital Institute of Pediatrics, Beijing, China）

Objective To study the relationship between metabolic syndrome and target organ damage (TOD) of essential hypertension in children.
Methods 165 children older than 10 years were retrospectively enrolled who were diagnosed as essential hypertension. All children were divided into group with metabolic syndrome (Group A) and group without metabolic syndrome (Group B). All clinical data were collected and analyzed using statistic methods.
Results In all recruited adolescents, 58 patients were diagnosed as metabolic syndrome with the rate of 35.2%. The incidence of target organ damage in Group A was significantly higher than Group B (*P*< 0.01）. The incidence of cardiovascular damage,especially left ventricular hypertrophy (LVH) in Group A was much higher than Group B (*P*<0.001,*P*<0.05）. The incidence of target organ damage in patients with impaired glucose tolerance was significantly higher than that of patients without those disorders (*P<0.05*). In particular, the high incidence of cardiac damage was significantly higher in the former (*P* <0.001). According to the results of multiple factors Logistic regression analysis, metabolic syndrome and elevated average diastolic pressure were independent risk factors for target organ damage (OR=3.538, OR=1.061).
Conclusions Essential hypertension complicated with metabolic syndrome in children had a much higher incidence of TOD than those without metabolic syndrome, especially cardiovascular damage. The examination of metabolic index is benefit for early diagnosis of target organ damage so as to take effective interference.

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[II-JCKP2-07] The effect of mother's social support on children with congenital heart disease and mother-child interaction on children's problem behavior.

○Ju Ryoung Moon[1,4,5,6], Jinyoung Song[1,2,4,5,6,7], June Huh[1,2,4,5,6,7], I-Seok Kang[1,2,4,5,6,7], Ji-Hyuk Yang[1,3,4,5,6,7], Tae-Gook Jun[1,3,4,5,6,7] （1.Grown-Up Congenital Heart Clinic, 2.Department of Pediatrics, 3.Department of Thoracic &Cardiovascular Surgery, 4.Cardiac Center, 5.Heart Vascular and Stroke Institute, 6.Samsung Medical Center, 7.Sungkyunkwan University School of Medicine）

Purpose: The purpose of this study was to examine the effect of mother’ s social support on children with congenital heart disease and mother-child interaction on children’ s problem behavior.
Methods: The data was collected from the outpatient clinic of cardiac disease at the Samsung Medical Center. 250 children with congenital heart disease, aged 4-6 years old, and their mothers were recruited for the study. The home environment, activities and cognitive stimulation questionnaire for mother-child

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interaction, social support and Korea-child behavior checklist were assessed and analyzed using Pearson' s correlations, stepwise multiple regressions in SPSS 22.0

Results: There were significant relationships found between age, education of mother' s demographic background, mother social support, mother-child interaction, and children' s problem behavior. Furthermore, the mother' s demographic background and mother social support indicated effective variance on mother-child interaction. Lastly, friends and colleagues of the mother and mother-child interaction were emphasized on effective variances on child problem behavior.

Conclusion : This study suggests that the mother' s social support system and environment has significant implications and should be further examined to better reduce and prevent problem behaviors in children.

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[II-JCKP2-08] Changes of content of circulating endothelial cells and endothelial progenitor cells in patients with primary hypertension in peripheral blood

○Lin Shi, Yuanyuan Zhang, Yao Lin, Yang Liu （Department of Cardiology,Capital Institute of Pediatrics, Beijing, China）

Background The key mechanism of target organ damage (TOD) in primary hypertension is vascular endothelial dysfunction. Circulating endothelial cells (CECs) and endothelial progenitor cell (EPCs) count and function testing was reported a new method for endothelial function and estimation.

Objectives To observe the concentration of CECs & EPCs in children with primary hypertension and their changes before and after the treatment.

Methods 64 cases with primary hypertension and 30 cases of healthy children were enrolled, who were divided into healthy children group (n = 30), pre-hypertension group (PHP) (n = 30), and high blood pressure（ HP） group (n = 64). Clinical data were collected and recorded. CECs and EPCs counts were measured by flow cytometry. Data were analysis by software SPSS 19.0.

Result CECs counts in PHP and HP group were higher than that of healthy group (P<0.05), while EPCs was significantly lower (P<0.05). However, there was no significant difference between PHP and HP group（P >0.05）. Meanwhile, there was no significant difference between stage 1 and 2 （P>0.05）.CECs counts in children with TOD was significantly higher than those without TOD (P<0.05), while EPCs was significantly lower (P<0.05). After 6-month treatment of diet and exercise, CECs counts in children with TOD was significantly lower than before， while EPCs was not significantly different.

Conclusions There were changes of CECs and EPCs counts in children with primary hypertension, especially in children with TOD. Diet and exercise may be helpful for the improvement of endothelial function.

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[II-JCKP2-09] Role of Cardiopulmonary Exercise Testing for Prediction of Pregnancy Outcome in Women with

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Congenital Heart Disease

○Midori Fukuyama, Hideo Ohuchi, Chizuko Kamiya, Mitsuhiro Fujino, Kenichi Kurosaki, Jun Yoshimatsu （National Cerebral and Cardiovascular Center, Japan）

Background: Most women with congenital heart disease (WCHD) are reaching reproductive adulthood and the pregnancy-associated issues have become one of major clinical practices in a field of WCHD. Prediction of pregnancy outcome is important and helpful to guide clinicians to care of WCHD. In this regard, NYHA may determine the outcome, there have been no objective criteria for the safer pregnancy in WCHD.
Purpose: To identify major risk factors and determine referral cardiovascular variables during cardiopulmonary exercise testing (CPX) in WCHD.
Method and Results: We retrospectively reviewed pregnancy outcomes of 68 WCHDs and those compared with NYHA, CPX-derived variables (heart rate [bpm], oxygen uptake, systolic blood pressure [SBP, mmHg] at peak exercise and clinically relevant arrhythmia during CPX(Ex-Arr)).17 maternal cardiac and 26 neonatal events occurred. All variables were associated with maternal cardiac and neonatal events ($p<0.05$-0.001). Of these, peak SBP and Ex-Arr were the independent determinants of the maternal (odds ratio [OR]:0.96, 95% confidence interval [CI]: 0.92-0.99, $p<0.05$ for peak SBP) and neonatal events (OR: 0.95, 95%CI: 0.91-0.99,$p<0.01$ for peak SBP, and OR: 21.2, 95%CI: 2.1-559,$p<0.01$ for Ex-Arr), except for Ex-Arr for maternal events. The cutoff value of SBP for maternal and neonatal events was 150 and 154.
Conclusion: All major CPX-derived variables, especially, peak SBP and Ex-Arr, can predict adverse outcome during pregnancy in WCHD. Peak SBP ≥ 150 without Ex-Arr could be a reliable reference value for safer pregnancy outcome in WCHD.

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[II-JCKP2-10] Myocardial Perfusion Abnormality by SPECT Imaging Correlates with Systemic Ventricular Property in Adults with Congenital Heart Disease

○Mitsuhiro Fujino, Hideo Ouch, Yosuke Hayama, Tohru Iwata, Jun Negishi, Aya Miyazaki, Etsuko Tsuda, Kenichi Kurosaki, （Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan）

【 Background】 The clinical role of myocardial perfusion imaging by single-photo emission computed tomography (SPECT-MPI) remains unclear in adults with congenital heart disease (ACHD).
【 Purpose】 This study was to clarify the clinical significance of SPECT-MPI for evaluating the mechanical function and electrical property of the systemic ventricle (SV) in ACHD patients.
【 Methods and Results】 Between August 2014 and February 2016, resting SPECT-MPI was performed in 95 ACHD patients (age: 31±10 years, biventricular physiology (BV) in 69 and Fontan physiology (Fontan) in 26). We developed MPI defect score (%D: %) as an index of severity of the SV myocardial perfusion abnormality and compared the index with plasma levels of high-sensitive troponin T (hsTnT), brain natriuretic peptide (BNP), end-diastolic volume index and ejection fraction of the SV (EF), QRS duration, and peak oxygen uptake (p VO2). In all ACHD patients, %D was 8.7±8.4. %D was independently correlated positively with QRS duration ($p<0.05$) and negatively with EF ($p<0.001$) and pVO2 ($p<0.05$),

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whereas %D was not correlated with hsTnT or BNP. These associations remained significant in both ACHD patients with BV ($p<0.05$-0.001) and Fontan ($p<0.05$), except for an association of %D with EF in Fontan.

【Conclusion】 SPECT-PMI has a significant role for providing us vital information on SV myocardial perfusion abnormality which is closely associated with SV mechano-electrical property in ACHD patients regardless of the hemodynamic pathophysiology.

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