# Sat. Jul 8, 2017

## ROOM 3

JCK Oral

### JCK Oral 1 (II-JCKO1)

### Basic/New Insights

Chair:Hiryoyuki Yamagishi(Department of Pediatrics, Keio University School of Medicine, Japan)
Chair:Yimin Hua(Department of Pediatric Cardiology, West China Second University Hosiptal, China)
Chair:Chung Il Noh(Department of Pediatrics, Seoul National University Hospital, Korea)
8:30 AM - 9:20 AM ROOM 3 (Exhibition and Event Hall Room 3)

[II-JCKO1-01] The Failing Right Heart in Congenital Heart Disease: New Mechanistic Insights, Echocardiographic Modalities Assessment and Management Strategies

○Yiu Fai Cheung (Department of Paediatrics and Adolescent Medicine, LKS Faculty of Medicine, The University of Hong Kong, Hong Kong)

8:30 AM - 9:20 AM

[II-JCKO1-02] Phenotype-Genotype correlations in the fetal patients with left ventricular noncompaction

○Keiichi Hirono[1], Asami Takasaki[1], Mako Okabe[1], Naruaki Miyao[1], Hideyuki Nakaoka[1], Kazuyoshi Saito[1], Sayaka Ozawa[1], Yukiko Hata[2], Naoki Nishida[2], Naoki Yoshimura[3], Fukiko Ichida[1] (1.Department of Pediatrics, Graduate School of Medicine, University of Toyama, Japan 2.Department of Legal Medicine, Graduate School of Medicine, University of Toyama, Japan 3.1st Department of Surgery, Graduate School of Medicine, University of Toyama, Japan)

8:30 AM - 9:20 AM

[II-JCKO1-04] Placental P-glycoprotein inhibition enhances susceptibility toDi(2-ethyhexyl)phthalateinduced cardiac malformations in mice

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

8:30 AM - 9:20 AM

[II-JCKO1-05] Roles of Tbx4 for pulmonary vascular development

○Keiko Uchida[1,2], Yu Yoshida[1], Reina Ishizaki[1], Akimichi Shibata[1], Kazuki Kodo[1], Takatoshi Tsuchihashi[1,3], Jun Maeda[1], Hiroyuki Yamagishi[1] (1.Department of Pediatrics, Keio University School of Medicine, Tokyo, Japan, 2.Health Center, Keio University, Kanagawa, Japan, 3.Department of Pediatrics, Kawasaki Municipal Hospital, Kanagawa, Japan)

8:30 AM - 9:20 AM

JCK Oral

### JCK Oral 2 (II-JCKO2)

### Fetal and Neonatal Cardiology

Chair:Noboru Inamura(Department of Pediatrics, Kindai University, Faculty of Medicine, Japan)
Chair:Xupei Huang(Department of Pediatric Cardiology, Guangdong General Hospital, China)
Chair:June Huh(Department of Pediatrics, Sungkyunkwan University School of Medicine, Heart Vascular Stroke Institute, Samsung Medical Center, Korea)
9:20 AM - 10:10 AM ROOM 3 (Exhibition and Event Hall Room 3)

[II-JCKO2-01] Sensitivity and positive predictive value of the fetal echocardiographic parameters to predict simple or complex coarctation of the aorta

○Mio Taketazu[1,2], Hirotaka Ishido[1], Sorachi Shimada[2], Yoichi Iwamoto[1], Satoshi Masutani[1], Hideaki Senzaki[1] (1.Department of Pediatric Cardiology, Saitama Medical Center, Saitama Medical University, Saitama, Japan, 2.Department of Pediatrics, Asahikawa Kosei General Hospital, Hokkaido, Japan)

9:20 AM - 10:10 AM

[II-JCKO2-03] Clinical research on fetal bradycardia

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

9:20 AM - 10:10 AM

[II-JCKO2-04] Basic Fetal Echocardiography

○Sin-weon Yun (Department of Pediatric Cardiology, Chung-Ang University Hospital, Korea )

9:20 AM - 10:10 AM

[II-JCKO2-05] A retrospective analysis to identify the

factors influencing parental decisions in pregnancies with fetal cardiac anomalies

○Lin Wu[1], Luming Sun[2] (1.Department of Cardiology, Children's Hospital of Fudan University, Shanghai-City, China, 2.Fetal Medicine Unit, Shanghai First Maternity and Infant Hospital, Tongji University School of Medicine, Shanghai-City, China)

9:20 AM - 10:10 AM

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JCK Oral

## JCK Oral 3 (II-JCKO3)

## Cardiac Surgery

Chair:Tetsuya Kitagawa(Department of Cardiovascular Surgery, Institute of Biomedical Sciences, Tokushima University Graduate School, Japan)
Chair:Fen Li(Department of Cardiology, Shanghai Children's Medical Center, China)
Chair:Tae Gook Jun(Department of Thoracic and Cardiovascular Surgery Sungkyunkwan University School of Medicine, Heart Vascular Stroke Institute, Samsung Medical Center,Korea)
1:50 PM - 3:20 PM ROOM 3 (Exhibition and Event Hall Room 3)

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[II-JCKO3-01] Coronary Morbidity after Arterial Switch Operation

○Woong Han Kim (Seoul National University, Seoul, Korea)

1:50 PM - 3:20 PM

[II-JCKO3-02] The necessity of TV repair with PVR in repaired TOF

○Jinyoung Song (Department of Pediatrics, Samsung Medical Center , Korea)

1:50 PM - 3:20 PM

[II-JCKO3-03] Surgical Repair of Congenital Mitral Valve Disease in Pediatric Population

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

1:50 PM - 3:20 PM

[II-JCKO3-04] Application of a simplified hand-sewn trileaflet valved conduit in RVOTreconstruction as an alternative for bovine jugular vein graft

○Huifeng Zhang, Bing Jia (Cardiovascular Center,Children's Hospital of Fudan University,Shanghai,China)

1:50 PM - 3:20 PM

[II-JCKO3-05] Mid-term Outcomes of Survival and Quality of Life in Children with Complex Congenital Heart Diseases after atrioventricular valvuloplasty in China

○Li Xiang, Yiwei Liu, Yuan Huang, Hao Zhang (Center of Pediatric Cardiac Surgery,National Center for Cardiovascular Diseases and Fuwai Hospital, Chinese Academy of Medical Sciences, Peking Union Medical College, Beijing, China)

1:50 PM - 3:20 PM

[II-JCKO3-06] A Novel Arterial Cannulation Technique for Selective Regional Cerebral Perfusion in Aortic Arch Surgery in Infants

○Hyungtae Kim[1], Si Chan Sung[1], Kwang Ho Choi[1], Hyoung Doo Lee[2], Geena Kim[2], Hoon Ko[2] (1.Department of Thoracic and Cardiovascular Surgery, Pusan National University Yangsan Hospital, Korea, 2.Department of Pediatrics, Pusan National University Yangsan Hospital, Korea)

1:50 PM - 3:20 PM

[II-JCKO3-07] Totally thoracoscopic without robotic assistance in cardiac surgery: the initial experience in Viet Nam and future perspectives

○Le Ngoc Thanh, Nguyen Cong Huu, Dang Quang Huy (Cardiovascular Center, E Hospital, Hanoi, Vietnam)

1:50 PM - 3:20 PM

[II-JCKO3-08] A study on the optimal extra-cardiac conduit size by comparison of exercise capacity according to conduit size in patients with Fontan circulation.

○Sang-Yun Lee[1], Seong-Ho Kim[1], So-Ick Jang[1], Soo-Jin Park[1], Eun-Young Choi[1], Hye-Won Kwon[1],Ji-Seok Bang[1], Ja-Kyung Yoon[1], Chang-Ha Lee[2], Eun-Seok Choi[2], Sung-Kyu Cho[2], (1.Department of Pediatrics, Sejong General Hospital, Bucheon-si, Korea, 2.Department of Cardiovascular and Thoracic Surgery, Sejong General Hospital, Bucheon-si, Korea)

1:50 PM - 3:20 PM

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JCK Oral

## JCK Oral 4 (II-JCKO4)

## Kawasaki Disease/General Cardiology 1

Chair:Masahiro Ishii(Department of Pediatrics, Kitasato

University School of Medicine, Kanagawa, Japan)
Chair:Min Huang(Shanghai Children's Hospital, China)
Chair:Seong-Ho Kim(Department of Pediatrics, Sejong General Hospital, Korea)
3:30 PM - 4:20 PM ROOM 3 (Exhibition and Event Hall Room 3)

[II-JCKO4-01] Severity assessment of coronary artery aneurysms in Kawasaki disease using the internal diameter Z-score

○Masaru Miura[1,2], Tohru Kobayashi[2], Naoya Fukushima[1,2], Taichi Kato[2], Shinya Shimoyama[2], Kenji Suda[2], Shigeto Fuse[2], Jun Maeda[2], Tsutomu Saji[2] (1.Department of Cardiology, Tokyo Metropolitan Children's Medical Center, Japan 2.The Z-Score Project 2nd Stage Study (ZSP2) Group, Japan)

3:30 PM - 4:20 PM

[II-JCKO4-02] Identification of susceptibility genes associated with Kawasaki disease by targeted enrichment of genomic region sequencing technique

○Danying Zhu[1], Han Zhang[1], Sirui Song[1], Lijian Xie[1], Feng Qiu[2], Jing Yang[2],Tingting Xiao[1], Min Huang[1], (1.Department of Cardiology,Shanghai Children's Hospital Affiliated to Shanghai Jiaotong University,Shanghai,China, 2.Shanghai Center for Bioinformation Technology)

3:30 PM - 4:20 PM

[II-JCKO4-03] Kawasaki shock syndrome complicated with macrophage activation syndrome

○Yao Lin, Lin Shi, Yanjun Deng, Yang Liu, Ping Lu (Department of Cardiology,Capital Institute of Pediatrics,Beijing,China)

3:30 PM - 4:20 PM

[II-JCKO4-04] The change of serum inflammatory meditators in 40 children with Kawasaki disease

○Wang Yun[1], Wang Yang[2], Ji Xiao Dan[2], Zhou Nan[3] (1.Department of Pediatrics, Beijing New Century Women's and Children's Hospital, Beijing, China, 2.New Century Beijing Children's Hospital, P.R. of China, 3.Beijing Children's Hospital, P.R. of China)

3:30 PM - 4:20 PM

[II-JCKO4-05] Evaluation of coronary vascular destruction by Optical Coherence Tomography and coronary artery diameter in acute phase of Kawasaki disease

○Ryuji Fukazawa, Yoshiaki Hashimoto, Makoto Watanabe, Kanae Tsuno, Koji Hashimoto, Miharu Akao, Mitsuhiro Kamisago, Yasuhiro Katsube (Department of Pediatrics, Nippon Medical School, Japan)

3:30 PM - 4:20 PM

JCK Oral

## JCK Oral 5 (II-JCKO5)
## Catheter Intervention

Chair:Satoshi Yazaki(Division of Pediatric Cardiology, Sakakibara Heart Institute, Tokyo, Japan)
Chair:Hong Gu(Department of Pediatric Cardiology, Beijing Anzhen Hospital, China)
Chair:Young-Hwue Kim(Department of Pediatric Cardiology, Congenital Heart Disease Center, Asan Medical Center, Korea)
4:20 PM - 5:10 PM ROOM 3 (Exhibition and Event Hall Room 3)

[II-JCKO5-01] New self-expandable percutaneous pulmonary valve implantation using knitted nitinol-wire stent mounted with a tri-leaflet porcine pericardial valve

○Gi Beom Kim, Mi Kyung Song, Eun Jung Bae, Chung Il Noh (Department of Pediatrics, Seoul National University Children's Hospital, Seoul, South Korea)

4:20 PM - 5:10 PM

[II-JCKO5-02] Stent of PDA as an alternative to Blalock-Taussig Shunt: single center experience

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

4:20 PM - 5:10 PM

[II-JCKO5-03] Tricuspid valve annulus Z score before and after transcatheter closure of atrial septal defect

○ChunMin Fu[1], Ming-Tai Lin[2], Jou-Kou Wang[2] (1.Department of Pediatrics, National Taiwan University Hospital, Hsinchu Branch, Taiwan, 2.Pediatric Cardiology, Department of Pediatrics, National Taiwan University Children's Hospital, Taiwan)

4:20 PM - 5:10 PM

[II-JCKO5-04] Hybrid pulmonary vein stenting in the patients with refractory to surgical pulmonary vein stenosis repair

○JaKyoung Yoon[1], Mi Kyung Song[2], Gi Beom Kim[2], Eun Jung Bae[2], Chung Il Noh[2], Jae Gun Kwak[3],Woong Han Kim[3], Jeong Ryul Lee[3], （1.Department of Pediatrics, Sejong General Hospital, Bucheon, South Korea, 2.Department of Pediatrics, Seoul National University Children's Hospital, Seoul, South Korea, 3.Department of Thoracic and Cardiovascular Surgery, Seoul National University Children's Hospital, Seoul, South Korea）
4:20 PM - 5:10 PM

[II-JCKO5-05] A Hybrid Biventricular CRT for a 6-year-old patient with heart failure after interventional closure of ventricular septal defect

○Yiwei Chen, Fen Li, Wei Ji, Jinjin Wu, Lijun Fu, Jie Shen,Shumin Wang, （Department of Cardiology, Shanghai Children's Medical Center Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai, PR China）
4:20 PM - 5:10 PM

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JCK Oral

## JCK Oral 6 (II-JCKO6)

## Long-term Outcome/ Heart Failure/Arrhythmia

Chair:Yoshiki Mori(Department of Pediatric Cardiology, Seirei Hamamatsu General Hospital, Japan)
Chair:Zhiwei Zhang(Department of Pediatric Cardiology, Guangdong General Hospital, China)
Chair:Si Chan Sung(Department of Thoracic and Cardiovascular Surgery, Pusan National University Yangsan Children's Hospital , Korea)
5:10 PM - 6:00 PM ROOM 3 (Exhibition and Event Hall Room 3)

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[II-JCKO6-01] Assessment of limited athletic participation school age after operation for severe congenital heart disease based on cardiovascular disease screening

○Satoru Iwashima[1], Satoshi Ueda[2], Keisuke Sato[1], Yasuhiko Tanaka[3] （1.Deaprtment of Pediatrics Cardiology, Chutoen General Medical Center, Kakegawa city, Japan, 2.Subcommittee for Discussing School Heart Examination Results of the Shizuoka Prefecture Medical Association, Japan, 3.Department of Pediatric Cardiology, Shizuoka Children's Hospital, Japan）
5:10 PM - 6:00 PM

[II-JCKO6-04] The fate of aortic valve after doubly commiitted juxtaarterial ventricular septal defect repair

○Hanna Jung, Young Ok Lee, Joon Yong Cho （Department of Thoracic and Cardiovascular Surgery, Kyungpook National University Hospital Korea, republic of Korea）
5:10 PM - 6:00 PM

[II-JCKO6-05] The outcome of junctional ectopic tachycardia following repair of congenital heart defects

○Yaping Mi, Bing Jia, Yonghao Gui （Cardiovascular Center, Children's Hospital, Fudan University, Shanghai, China）
5:10 PM - 6:00 PM

## Sun. Jul 9, 2017

## ROOM 3

JCK Oral

### JCK Oral 7 (III-JCKO7)

### Kawasaki Disease/General Cardiology 2

Chair:Mamoru Ayusawa(Department of Pediatrics and Child Health Nihon University school of Medicine & Itabashi Hospital, Japan)
Chair:Lucy Youngmin Eun(Department of Pediatric Cardiology, Yonsei University, Seoul, Korea)
10:15 AM - 11:05 AM ROOM 3 (Exhibition and Event Hall Room 3)

[III-JCKO7-01] Experience of recurrent Kawasaki disease complicated by giant aneurysms showing only two of the main symptoms

○Chika Nakamura[1], Yukiko Kawazu[1], Shinji Kaichi[2], Etsuko Tsuda[3] (1.Department of Pediatrics, Toyonaka Municipal Hospital, Toyonaka-City, Osaka, Japan, 2.Department of Pediatric Cardiology, Hyogo Prefectural Amagasaki General Medical Center, Hyogo, Japan, 3.Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan)

10:15 AM - 11:05 AM

[III-JCKO7-02] Clinical analysis of hospitalized children with Kawasaki disease based on E - Science model, single center registry from 2009 to 2016

○Sirui Song, Danying Zhu, Min Huang, Lijian Xie, Tingting Xiao (Department of Cardiology,Shanghai Children's Hospital Affiliated to Shanghai Jiaotong University,Shanghai,China)

10:15 AM - 11:05 AM

[III-JCKO7-03] Kawasaki Disease with Atypical Presentation Masquerading as Severe Infection: a 10-year retrospective review in a Tertiary Hospital in Hong Kong

○Chi Yu Dennis Au, Nai Chung Fong, Cheuk Man Ronald Fung, Wai Yau Daniel Mak, Wai Lim Yiu, Yu Ming Fu (Department of Pediatrics and Adolescent Medicine, Princess Margaret Hospital, Hong Kong SAR, China)

10:15 AM - 11:05 AM

[III-JCKO7-04] Coronary Artery Bypass Grafting in Children with Severe Coronary artery lesions after Kawasaki Disease

○Wenbo Zhang, Ming Ye, Bing Jia, Fang Liu, Lin Wu (Cardiovascular Center, Children's Hospital of Fudan University, Shanghai , China)

10:15 AM - 11:05 AM

[III-JCKO7-05] Risk factors and implications of progressive coronary dilatation in children with Kawasaki disease

○Ming-Tai Lin, Ming-Yu Liu, Chia-Hui Wu, Jou-Kou Wang, Mei-Hwan Wu (Department of Pediatrics, National Taiwan University Hospital, Taiwan)

10:15 AM - 11:05 AM

JCK Oral

### JCK Oral 8 (III-JCKO8)

### Cardiovascular Imaging

Chair:Keisuke Satou(Department of Cardiology Shizuoka Children Hospital, Japan)
Chair:Seong-Ho Kim(Department of Pediatrics, Sejong General Hospital, Korea)
11:05 AM - 11:55 AM ROOM 3 (Exhibition and Event Hall Room 3)

[III-JCKO8-01] Morphological and clinical spectrum of infantile scimitar syndrome

○Lin Wu, Fang Liu (Department of Cardiology, Children's Hospital of Fudan University, Shanghai-City, China)

11:05 AM - 11:55 AM

[III-JCKO8-02] New fusion imaging - VesselNavigator in CHD interventions

○Seong Ho Kim, Su Jin Park, Hye Won Kwon, Sang Yun Lee, Ji Seok Bang, Eun Young Choi, So Ick Jang, Ja Kyung Yun (Department of Pediatrics, Sejong General Hospital, Korea)

11:05 AM - 11:55 AM

[III-JCKO8-03] Cardiac Mechanics in Children post Percutaneous Transcatheter Closure of Perimembranous Ventricular Septal Defect

○Lijian Xie, Xunwei Jiang, Yun Li, Min Huang, Tingting Xiao (Department of Cardiovascular, Shanghai Children's Hospital, Shanghai Jiaotong University, Shanghai,

China）

11:05 AM - 11:55 AM

[III-JCKO8-05] Evaluation of tricuspid valve displacement in repaired tetralogy of Fallot using feature tracking magnetic resonance program

○Akio Inage[1], Naokazu Mizuno[2]（1.Division of Pediatric Cardiology, Sakakibara Heart Institute, Japan, 2.Division of Radiology, Sakakibara Heart Institute, Japan）

11:05 AM - 11:55 AM

JCK Oral

# JCK Oral 1 (II-JCKO1)

## Basic/New Insights

Chair:Hiryoyuki Yamagishi(Department of Pediatrics, Keio University School of Medicine, Japan)
Chair:Yimin Hua(Department of Pediatric Cardiology, West China Second University Hosiptal, China)
Chair:Chung Il Noh(Department of Pediatrics, Seoul National University Hospital, Korea)
Sat. Jul 8, 2017 8:30 AM - 9:20 AM ROOM 3 (Exhibition and Event Hall Room 3)

### [II-JCKO1-01] The Failing Right Heart in Congenital Heart Disease: New Mechanistic Insights, Echocardiographic Modalities Assessment and Management Strategies

○Yiu Fai Cheung （Department of Paediatrics and Adolescent Medicine, LKS Faculty of Medicine, The University of Hong Kong, Hong Kong）

8:30 AM - 9:20 AM

### [II-JCKO1-02] Phenotype-Genotype correlations in the fetal patients with left ventricular noncompaction

○Keiichi Hirono[1], Asami Takasaki[1], Mako Okabe[1], Naruaki Miyao[1], Hideyuki Nakaoka[1], Kazuyoshi Saito[1], Sayaka Ozawa[1], Yukiko Hata[2], Naoki Nishida[2], Naoki Yoshimura[3], Fukiko Ichida[1] （1.Department of Pediatrics, Graduate School of Medicine, University of Toyama, Japan 2.Department of Legal Medicine, Graduate School of Medicine, University of Toyama, Japan 3.1st Department of Surgery, Graduate School of Medicine, University of Toyama, Japan）

8:30 AM - 9:20 AM

### [II-JCKO1-04] Placental P-glycoprotein inhibition enhances susceptibility toDi(2-ethyhexyl)phthalateinduced cardiac malformations in mice

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

8:30 AM - 9:20 AM

### [II-JCKO1-05] Roles of Tbx4 for pulmonary vascular development

○Keiko Uchida[1,2], Yu Yoshida[1], Reina Ishizaki[1], Akimichi Shibata[1], Kazuki Kodo[1], Takatoshi Tsuchihashi[1,3], Jun Maeda[1], Hiroyuki Yamagishi[1] （1.Department of Pediatrics, Keio University School of Medicine, Tokyo, Japan, 2.Health Center, Keio University, Kanagawa, Japan, 3.Department of Pediatrics, Kawasaki Municipal Hospital, Kanagawa, Japan）

8:30 AM - 9:20 AM

8:30 AM - 9:20 AM (Sat. Jul 8, 2017 8:30 AM - 9:20 AM ROOM 3)

# [II-JCKO1-01] The Failing Right Heart in Congenital Heart Disease: New Mechanistic Insights, Echocardiographic Modalities Assessment and Management Strategies

○Yiu Fai Cheung （Department of Paediatrics and Adolescent Medicine, LKS Faculty of Medicine, The University of Hong Kong, Hong Kong）

Normal functioning of the subpulmonary and systemic right ventricles is important in congenial heart patients. Recent studies have provided mechanistic insights on progressive failure of the right heart, which include possible roles of altered right ventricular molecular remodeling response, altered extracellular matrix composition, cardiac apoptosis, microRNA, and genetic polymorphisms. New echocardiographic techniques have allowed quantification beyond assessment of right ventricular volumes and ejection fraction and enabled direct interrogation of right atrial and ventricular deformation. The novel vector flow mapping may further allow bedside quantification of pulmonary regurgitation. Management of right ventricular dysfunction in the setting of congenital heart disease remains challenging. Conventional heart failure medications have been used with disappointing results. Possible beneficial effects of vasopressors have been shown in the management of acute right ventricular failure in decompensated patients. Cardiac resynchronization therapy has shown some promise although long-term data are lacking. The optimal timing of pulmonary valve replacement in patients with repaired tetralogy and the role of tricuspid valve repair in adult patients with a failing systemic right ventricle to ameliorate right ventricular volume load remains to be subjects of debate. Further understanding of pathogenesis of progressive right ventricular dysfunction is important for discovery of target-specific new therapies.

8:30 AM - 9:20 AM (Sat. Jul 8, 2017 8:30 AM - 9:20 AM ROOM 3)

# [II-JCKO1-02] Phenotype-Genotype correlations in the fetal patients with left ventricular noncompaction

○Keiichi Hirono[1], Asami Takasaki[1], Mako Okabe[1], Naruaki Miyao[1], Hideyuki Nakaoka[1], Kazuyoshi Saito[1], Sayaka Ozawa[1], Yukiko Hata[2], Naoki Nishida[2], Naoki Yoshimura[3], Fukiko Ichida[1] （1.Department of Pediatrics, Graduate School of Medicine, University of Toyama, Japan 2.Department of Legal Medicine, Graduate School of Medicine, University of Toyama, Japan 3.1st Department of Surgery, Graduate School of Medicine, University of Toyama, Japan）

Background:
Left ventricular noncompaction (LVNC) is a hereditary cardiomyopathy and associated with high morbidity and mortality, but the genetic background has not been fully evaluated. The aim of the present study was to identify the genetic background using next-generation sequencing (NGS) and to identify genotype-phenotype correlations in fetal patients with LVNC.
Methods:
We screened 73 genes associated with a cardiomyopathy in 20 fetal patients (11 males and 9 females) with LVNC for mutations by next generating sequencing (NGS). We compared the clinical features, anatomical properties and long-term prognosis between fetal patients and 111 other age patients with

LVNC.
Results:
The age at diagnosis ranged at 21 to 36 week' s gestation (median: 29 week' s gestation). Seven patients had a family history. Seventeen patients had congestive heart failure (CHF) and 5 patients had arrhythmias. Associated congenital heart diseases were identified in 5 patients. Nine patients died and 3 patients had intrauterine death or termination of pregnancy. Fourteen pathogenic mutations were found among 7 genes in 12 patients; 7 mutations in *MYH7* gene and 8 were novel. The *MYH7* group presented with lower age at onset and higher prevalence of congenital heart defects than that without *MYH7* mutations. The fetal patients had more frequency of positive for *MYH7* gene mutations and higher mortality than other age patients. The multivariable proportional hazards model showed that fetal patients and CHF at diagnosis were independent risk factors for death in all LVNC patients.
Conclusions:
The present study was the first report focused on genotype-phenotype relationships in fetal patients with LVNC using NGS. *MYH7* gene mutations can be used to predict the risk of other congenital heart in fetal LVNC patients and might have a pivotal role during maturation of heart in the fetus.

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8:30 AM - 9:20 AM (Sat. Jul 8, 2017 8:30 AM - 9:20 AM ROOM 3)

## [II-JCKO1-04] Placental P-glycoprotein inhibition enhances susceptibility toDi(2-ethyhexyl)phthalateinduced cardiac malformations in mice

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

**Purpose:**This study aims to explore whether inhibition of placental P-gp function with verapamil could enhance susceptibility to DEHP induced cardiac malformations in mice or not. **Methods:** The pregnant C57BL mice were randomized into the vehicle group (n=10), the DEHP group (n=20, 1g/Kg), the verapamil group (n=10, 3mg/Kg) and the DEHP+verapamil group (n=20). Pregnant dams in different groups received respective interventions by gavage once daily from E6.5-E14.5. Maternal weights were monitored everyday and samples were collected at E15.5. HE staining was used to examine fetal cardiac malformations. Fetal cardiac development-related genes (Nkx2.5/Gata4/Tbx5/Mef2c/Chf1) mRNA and protein expression were determined by quantitative real-timePCR (qRT-PCR) andwestern blot (WB), respectively. Maternal modality, maternal complete stillbirth/ abortion rates and fetal cardiac malformations rates were also calculated. **Results:** Maternal modality, maternal complete stillbirth/abortion rates and fetal cardiac malformations rates of DEHP+verapamil group were significantly higher than that of DEHP group, verapamil group and vehicle group. Compared with DEHP group, verapamil group and vehicle group, fetal cardiac Gata4/Mef2c/Chf1 expression was significantly down regulated in DEHP+verapamil group. There were no differences in above parameters between verapamil group and vehicle group.
**Conclusions:**Placental P-glycoprotein inhibition could enhance susceptibility to DEHP induced cardiac malformations in mice.

8:30 AM - 9:20 AM (Sat. Jul 8, 2017 8:30 AM - 9:20 AM ROOM 3)

# [II-JCKO1-05] Roles of Tbx4 for pulmonary vascular development

○Keiko Uchida[1,2], Yu Yoshida[1], Reina Ishizaki[1], Akimichi Shibata[1], Kazuki Kodo[1], Takatoshi Tsuchihashi[1,3], Jun Maeda[1], Hiroyuki Yamagishi[1] (1.Department of Pediatrics, Keio University School of Medicine, Tokyo, Japan, 2.Health Center, Keio University, Kanagawa, Japan, 3.Department of Pediatrics, Kawasaki Municipal Hospital, Kanagawa, Japan)

[Objectives] A T-box transcription factor, Tbx4, is involved in embryogenesis. Recently, it was reported that Tbx4 was expressed in the lung mesenchymal cells (LMC), possible precursors for pulmonary arterial endothelial cells (PAEC) and smooth muscle cells (PASMC). In addition, some microdeletions and mutations in the genome region of Tbx4 were detected in the patients with childhood-onset pulmonary arterial hypertension. The purpose of this study is to elucidate roles of Tbx4 for the development of the pulmonary vessels.
[Methods] We performed microarray analysis compared between CD31-positive cells sorted from the lung tissues at embryonic day (E) E14 and those at postnatal day (P) 2. Then we focused on Tbx4 as a molecule expressed highly at E14. The temporal expression pattern of Tbx4 in LMC was observed by qPCR. *In vitro* tube formation activity using Tbx4-knockdown LMC and BrdU incorporation assay using Tbx4-knockdown PASMC were examined.
[Results] Microarray showed the expression level of Tbx4 at E14 was significantly higher than that at P2. Interestingly, Tbx4 expression level in the LMC reached to the peak at E14-15 and later decreased. The knockdown of Tbx4 in LMC increased tube length in tube formation assay. In contrast the knockdown in the PASMC attenuated cell proliferation activities.
[Conclusions] Our results suggest that Tbx4 in the LMC may have roles for maintaining immaturity of precursors of pulmonary vascular cells and that the downregulation of Tbx4 expression after E14-15 may proceed their differentiation.

JCK Oral

# JCK Oral 2 (II-JCKO2)

## Fetal and Neonatal Cardiology

Chair:Noboru Inamura(Department of Pediatrics, Kindai University, Faculty of Medicine, Japan)
Chair:Xupei Huang(Department of Pediatric Cardiology, Guangdong General Hospital, China)
Chair:June Huh(Department of Pediatrics, Sungkyunkwan University School of Medicine, Heart Vascular Stroke Institute, Samsung Medical Center, Korea)
Sat. Jul 8, 2017 9:20 AM - 10:10 AM ROOM 3 (Exhibition and Event Hall Room 3)

### [II-JCKO2-01] Sensitivity and positive predictive value of the fetal echocardiographic parameters to predict simple or complex coarctation of the aorta

○Mio Taketazu[1,2], Hirotaka Ishido[1], Sorachi Shimada[2], Yoichi Iwamoto[1], Satoshi Masutani[1], Hideaki Senzaki[1] （1.Department of Pediatric Cardiology, Saitama Medical Center, Saitama Medical University, Saitama, Japan, 2.Department of Pediatrics, Asahikawa Kosei General Hospital, Hokkaido, Japan）

9:20 AM - 10:10 AM

### [II-JCKO2-03] Clinical research on fetal bradycardia

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

9:20 AM - 10:10 AM

### [II-JCKO2-04] Basic Fetal Echocardiography

○Sin-weon Yun （Department of Pediatric Cardiology, Chung-Ang University Hospital, Korea ）

9:20 AM - 10:10 AM

### [II-JCKO2-05] A retrospective analysis to identify the factors influencing parental decisions in pregnancies with fetal cardiac anomalies

○Lin Wu[1], Luming Sun[2] （1.Department of Cardiology, Children's Hospital of Fudan University, Shanghai-City, China, 2.Fetal Medicine Unit, Shanghai First Maternity and Infant Hospital, Tongji University School of Medicine, Shanghai-City, China）

9:20 AM - 10:10 AM

9:20 AM - 10:10 AM (Sat. Jul 8, 2017 9:20 AM - 10:10 AM ROOM 3)

## [II-JCKO2-01] Sensitivity and positive predictive value of the fetal echocardiographic parameters to predict simple or complex coarctation of the aorta

○Mio Taketazu[1,2], Hirotaka Ishido[1], Sorachi Shimada[2], Yoichi Iwamoto[1], Satoshi Masutani[1], Hideaki Senzaki[1] (1.Department of Pediatric Cardiology, Saitama Medical Center, Saitama Medical University, Saitama, Japan, 2.Department of Pediatrics, Asahikawa Kosei General Hospital, Hokkaido, Japan)

Objective: To assess the usefulness of the known echocardiographic parameters to identify fetal simple (S) or complex (C) coarctation of the aorta (CoA).
Methods: All fetuses without semilunar valve atresia or interrupted aortic arch were enrolled. The echocardiograms were retrospectively analyzed to assess the previously reported predictors: pulmonary valve / aortic valve diameter ratio (PV/AV) >=1.6, Z-score of the ascending aorta diameter (AA-Z) <= -1.5 (Gomez-Montes, 2013); Z-score of the isthmus diameter (IS-Z) <-2, the isthmus to ductal ratio (IS/DA) <0.74, isthmus flow disturbance, visualization of the posterior shelf (shelf) (Matsui, 2008). If the fetuses had one or more predictors, they were considered as suspected CoA. The sensitivity, the specificity, and the positive predictive value (PPV) of the each parameter were calculated to predict the postnatal CoA.
Results: Among 513 fetus with normal heart structure (group S), 16 had suspected CoA, and 5 had the postnatally confirmed CoA. Among 153 fetuses with a major heart defect (group C), 22 had suspected CoA, and 17 had the postnatally confirmed CoA. All of the confirmed CoA had one or more predictors. No baby without the suspected CoA had postnatal CoA. In total, “suspected CoA” had 100%-sensitivity and 98%- specificity and PPV of 58% in this study. All parameters had high specificities (98-99%), but sensitivities of individual parameters were various (45.5-90.9%). In group S, while “suspected CoA” had high sensitivity/specificity (100%/97.8%), its PPV was low (31.3%). IS-Z had high sensitivity of 100% but low PPV of 41.7%. The shelf had 100%-PPV, but its sensitivity remained low at 60%. In contrast, “suspected CoA” in group C had high sensitivity (100%) and high PPV (77.3%). Individually, IS-Z and IS/DA had high sensitivities (88.2% and 94.1%) and high PPVs (83.3% and 84.2%), respectively.
Conclusion: Specificities/sensitivities of these parameters could be high by using them combined. They also can prenatally detect simple CoA while the improvement of the PPV needs to be pursued. In contrast, these predictors provided an accurate prenatal diagnosis of the complex CoA.

9:20 AM - 10:10 AM (Sat. Jul 8, 2017 9:20 AM - 10:10 AM ROOM 3)

## [II-JCKO2-03] Clinical research on fetal bradycardia

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

**Obiective:** Investgate the disgnosis and prognosis of fetal bradycardia. **Methods:** With the determined of prenatal echocardiography and postnatal electrocardiogram, whileas the serum level of maternal anti-SSA/Ro and anti-SSB/La and status of maternal autoimmune diseases. **Results:** 47 cases of fetal bradycardia were enrolled, the gestational age were 18-34 weeks. Among them, there were 21 cases of III° AVB, 16 cases of sinus bradycardia, 8 cases of irregular bradycardia and 2 cases of I° AVB. Maternal autoantibodies positive were found in 26 mother; and to their fetuses, there were 17 cases of III° AVB, 5

cases of sinus bradycardia, 2 cases of irregular bradycardia. To the suffered fetuses, 17 cases were companied with cardiovascular malformation (12 cases of III° AVB, 3 cases of sinus bradycardia, and 2 cases of irregular bradycardia). The 2 cases of I° AVB (obviously prolonged A-V interval) in this study received prenatal dexamethasone therapy, halt the progress of heart block successfully and delivered two health babies. All the fetuses HR below 55bpm were terminated after prenatal doagnosis.
**Conclusion:**The prognosis of fetal bradycardia was poor, and some types of fetal bradycardia have closely relationship to connective tissue disease, especially maternal autoantibodies positive. Transplacental dexamethasone therapy for prolonged A-V interval may halt the progress of heart block and result good prognosis.

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9:20 AM - 10:10 AM (Sat. Jul 8, 2017 9:20 AM - 10:10 AM ROOM 3)

## [II-JCKO2-04] Basic Fetal Echocardiography

○Sin-weon Yun （Department of Pediatric Cardiology, Chung-Ang University Hospital, Korea ）

In recent years, marked advances in the technology, the field of prenatal diagnosis of congenital heart disease(CHD) is no longer unfamiliar. Understanding of antenatal diagnosis of the significant heart anomalies can be marked to improve neonatal outcome. Nevertheless, CHDs are the most frequently overlooked fields during fetal work-up, and thus may lead medicolegal, socioeconomic and psychological problems. Antenatal detection of cardiac anomalies is commonly made between 18th ~ 24th weeks of gestational age. Subtle clues in the early life may important key of diagnosis. Fetal sonographers and pediatric cardiologist need familiar with normal and abnormal fetal sequential axial scan. There are standard several transverse and sagittal views, Transverse views are easy to obtain and be standardized. Furthermore, a serial transverse section provides anatomic information. The other views parallel to the fetal long axis can be divided into coronal and longitudinal views. Coronal views are hardly possible to get or informative. It can always emphasize that serial axial scanning is important. Confirmed with 2D and check the flow pattern with color and Doppler, and try to find any great arteries discrepancies and isthmus hypoplasia. And confirm the normal pulmonary venous return use low velocity color Doppler. Some CHDs such as so-called right side obstructive lesions (such as pulmonary stenosis, tetralogy of the Fallot, pulmonary atresia series) and left sided obstructive lesions (such as critical aortic stenosis, hypoplastic left heart syndrome, coarctation of the aorta, interrupted aortic arch series) may progress into the gestational age, so if suspicious these anomalies, regular follow- up for progression is mandatory. And examiner’ s experience and acknowledge of the postnatal outcome may greatly influence to the counseling attitude and final fetal outcome. Some serious cardiac lesions may need emergency intervention just after delivery, so decide for delivery plan also crucial for outcome.

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9:20 AM - 10:10 AM (Sat. Jul 8, 2017 9:20 AM - 10:10 AM ROOM 3)

## [II-JCKO2-05] A retrospective analysis to identify the factors influencing parental decisions in pregnancies with fetal cardiac anomalies

○Lin Wu[1], Luming Sun[2] （1.Department of Cardiology, Children's Hospital of Fudan University, Shanghai-City, China, 2.Fetal Medicine Unit, Shanghai First Maternity and Infant Hospital, Tongji University School of Medicine, Shanghai-City, China）

**Objective** The aim of the study is to identify the impact of prenatal diagnosis of cardiac anomalies on parental decision of pregnancy termination in Chinese population in the setting of tertiary level diagnosis and consultation. **Methods** A total of 162 fetuses prenatally diagnosed with cardiac anomalies in our institute were retrospectively analyzed from January 2011 to December 2014. **Result** Of the 162 cases, the mean gestational age at diagnosis was 26.5 weeks (range from 17.4 to 39.5 weeks), and 24 fetuses (14.8%) were associated with major extra-cardiac malformations. Overall, 67 (41.4%) pregnancies were interrupted, while the rates of termination in mild, moderate and severe group of fetuses classified by severity of cardiac anomalies were 16.0%, 51.1%, and 76.2%, respectively, and that of fetuses associated with major extra-cardiac anomalies reached 79.2%. Multivariable logistic regression analysis identified that severity of cardiac anomalies (OR 9.001, p=0.000), presence of extra-cardiac anomalies (OR 3.801, p=0.000) and gestational age at diagnosis (OR 0.750, p=0.000) are three major factors contributing to the probability of pregnancy termination. **Conclusion** In China, the termination of pregnancy following prenatal diagnosis of cardiac anomalies is more frequent than that in the developed countries, mainly due to no restriction of gestational age for termination. The parents are more likely to opt for termination in the cases of more complex cardiac anomalies or in association with major extra-cardiac malformations.

JCK Oral

# JCK Oral 3 (II-JCKO3)

## Cardiac Surgery

Chair:Tetsuya Kitagawa(Department of Cardiovascular Surgery, Institute of Biomedical Sciences, Tokushima University Graduate School, Japan)
Chair:Fen Li(Department of Cardiology, Shanghai Children's Medical Center, China)
Chair:Tae Gook Jun(Department of Thoracic and Cardiovascular Surgery Sungkyunkwan University School of Medicine, Heart Vascular Stroke Institute, Samsung Medical Center,Korea)
Sat. Jul 8, 2017 1:50 PM - 3:20 PM ROOM 3 (Exhibition and Event Hall Room 3)

### [II-JCKO3-01] Coronary Morbidity after Arterial Switch Operation

○Woong Han Kim （Seoul National University, Seoul, Korea）
1:50 PM - 3:20 PM

### [II-JCKO3-02] The necessity of TV repair with PVR in repaired TOF

○Jinyoung Song （Department of Pediatrics, Samsung Medical Center , Korea）
1:50 PM - 3:20 PM

### [II-JCKO3-03] Surgical Repair of Congenital Mitral Valve Disease in Pediatric Population

○Bing Jia （Department of Children's Hospital ,FUDan University,Shanghai,China）
1:50 PM - 3:20 PM

### [II-JCKO3-04] Application of a simplified hand-sewn trileaflet valved conduit in RVOTreconstruction as an alternative for bovine jugular vein graft

○Huifeng Zhang, Bing Jia （Cardiovascular Center,Children's Hospital of Fudan University,Shanghai,China）
1:50 PM - 3:20 PM

### [II-JCKO3-05] Mid-term Outcomes of Survival and Quality of Life in Children with Complex Congenital Heart Diseases after atrioventricular valvuloplasty in China

○Li Xiang, Yiwei Liu, Yuan Huang, Hao Zhang （Center of Pediatric Cardiac Surgery,National Center for Cardiovascular Diseases and Fuwai Hospital, Chinese Academy of Medical Sciences, Peking Union Medical College, Beijing, China）
1:50 PM - 3:20 PM

### [II-JCKO3-06] A Novel Arterial Cannulation Technique for Selective Regional Cerebral Perfusion in Aortic Arch Surgery in Infants

○Hyungtae Kim[1], Si Chan Sung[1], Kwang Ho Choi[1], Hyoung Doo Lee[2], Geena Kim[2], Hoon Ko[2] （1.Department of Thoracic and Cardiovascular Surgery, Pusan National University Yangsan Hospital, Korea, 2.Department of Pediatrics, Pusan National University Yangsan Hospital, Korea）
1:50 PM - 3:20 PM

### [II-JCKO3-07] Totally thoracoscopic without robotic assistance in cardiac surgery: the initial experience in Viet Nam and future perspectives

○Le Ngoc Thanh, Nguyen Cong Huu, Dang Quang Huy （Cardiovascular Center, E

Hospital, Hanoi, Vietnam）

1:50 PM - 3:20 PM

## [II-JCKO3-08] A study on the optimal extra-cardiac conduit size by comparison of exercise capacity according to conduit size in patients with Fontan circulation.

○Sang-Yun Lee[1], Seong-Ho Kim[1], So-Ick Jang[1], Soo-Jin Park[1], Eun-Young Choi[1], Hye-Won Kwon[1],Ji-Seok Bang[1], Ja-Kyung Yoon[1], Chang-Ha Lee[2], Eun-Seok Choi[2], Sung-Kyu Cho[2], （1.Department of Pediatrics, Sejong General Hospital, Bucheon-si, Korea, 2.Department of Cardiovascular and Thoracic Surgery, Sejong General Hospital, Bucheon-si, Korea）

1:50 PM - 3:20 PM

1:50 PM - 3:20 PM (Sat. Jul 8, 2017 1:50 PM - 3:20 PM ROOM 3)

## [II-JCKO3-01] Coronary Morbidity after Arterial Switch Operation

○Woong Han Kim (Seoul National University, Seoul, Korea)

**Objective**: Coronary artery morbidity after an arterial switch operation causes subsequent reoperation. We investigated the freedom from reoperation, risk factors for reoperation, and results of reoperation.
**Methods**: Between Sep. 2003 and Dec. 2016, 79 consecutive patients who underwent an arterial switch operation and survived the early postoperative period were included. Preoperative characteristics and operative techniques were investigated in the risk factor analysis. The reoperation techniques and postoperative results were analyzed.
**Results**: There were no late deaths. Seven patients underwent reoperation due to coronary morbidity. Freedom from reoperation at 5 years and 10 years after the initial operation were 94.5% and 88.6%, respectively. Multivariate analysis revealed that a coronary artery between the great arteries and a high take-off coronary artery were significant risk factors for reoperation. Reoperation techniques included coronary artery ostium unroofing, cut-back angioplasty, and ostioplasty. No patients who underwent unroofing and cut-back angioplasty experienced complications during the median follow-up period of 36.7 months. However, 2 patients who underwent ostioplasty required an additional reoperation due to coronary artery restenosis.
**Conclusions**: A coronary artery between the great arteries and a high take-off coronary artery were significant risk factors for reoperation due to coronary artery stenosis following the arterial switch operation. Good reoperation results were observed using the unroofing and cut-back angioplasty techniques.

1:50 PM - 3:20 PM (Sat. Jul 8, 2017 1:50 PM - 3:20 PM ROOM 3)

## [II-JCKO3-02] The necessity of TV repair with PVR in repaired TOF

○Jinyoung Song (Department of Pediatrics, Samsung Medical Center , Korea)

Tricuspid regurgitation (TR) in repaired TOF has known to be functional due to right ventricular dilatation produced by chronic pulmonary regurgitation. There is a guideline that suggests pulmonary valve replacement when significant tricuspid regurgitation is progressive. Therefore, spontaneous improvement of TR could be expected after pulmonary valve replacement in repaired TOF but clinical outcomes were not sufficient. Studies about pulmonary valve replacement in repaired TOF revealed TR repair was performed concomitantly in 13-41% of patients. It is true that TR in repaired TOF is not only functional but also pathologic. Previous reports have shown that some patients with repaired TOF have tricuspid leaflet abnormalities or displacement after ventricular septal defect repair or ventricular pacing lead placement. It is important to differentiate the reasons of TR that make different treatment. In the era of percutaneous pulmonary valve replacement, the necessity of TR repair is getting more important. They recommended surgical insertion of pulmonary valve when TR should be repaired. And over we don' t have clear surgical indications of TR repair associated with repaired TOF. There is a guideline for TR repair when MV repair is to be expected. We reviewed TR mechanism in repaired TOF, the consequences of pulmonary valve replacement in repaired TOF and safety and confidence of TR repair in repaired TOF.

1:50 PM - 3:20 PM (Sat. Jul 8, 2017 1:50 PM - 3:20 PM ROOM 3)

## [II-JCKO3-03] Surgical Repair of Congenital Mitral Valve Disease in Pediatric Population

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

**Objective** Surgical treatment of congenital mitral valve disease is difficult in pediatric population. This article aims to explore diagnosis and surgical treatment of congenital mitral valve disease in pediatric population.**Methods** Between January 2008 and December 2015, 76 patients underwent mitral valve repair for congenital mitral valve disease. The average age of repair was 3.8±2.5 years (18 cases <2 years, 7 cases <1 years ). 21 patients had associated cardiac defects. In 52 patients with mitral regurgitation, surgical techniques include clefts closure(16 cases), commissuroplasty(16 cases), annuloplasty(12 cases), chordal shortening(8 cases), anterior/posterior leaflet extension(6 cases). In 24 patients with mitral stenosis , surgical techniques include supramitral ring excision(16 cases), commissures excision(6 cases), papillary muscles spliting(2 cases), chordal detachment and spliting (3 cases).**Results** There were 3 death in all patients (3.9%), 1 death in mitral regurgitation group (1.9%) and 2 death in mitral stenosis group(8.3%). Reoperation happened in 9 patients. The type of reoperation including mechanical valve replacement(3 cases,2 cases accept reoperation in early followup) and mitral valve repair(6 cases) . There were no death in reoperation patients.**Conclusions** Surgical repair of congenital mitral mitral valve disease is still facing a huge challenge. Compared to mitral regurgitation, the mortality in mitral stenosis is higher. Some patients need multiple operations, valvoplasty is the first consideration.

1:50 PM - 3:20 PM (Sat. Jul 8, 2017 1:50 PM - 3:20 PM ROOM 3)

## [II-JCKO3-04] Application of a simplified hand-sewn trileaflet valved conduit in RVOTreconstruction as an alternative for bovine jugular vein graft

○Huifeng Zhang, Bing Jia （Cardiovascular Center,Children's Hospital of Fudan University,Shanghai,China）

***Background:*** The Bovine jugular vein (BJV) graft for right ventricular outflow tract reconstruction (RVOT) is limited applied due to possible graft failure. In this study, we reported the clinical application of simplified hand-sewn trileaflet valved conduit as an alternative for BJV graft.

***Methods:*** We retrospectively included 68 patients underwent 73 conduits implantation including 22 new simplified hand-sewn trileaflet valved conduits (Group A) and 54 BJV grafts (Group B). For patients in Group A, a hand-sewn trileaflet valved conduit with valves made of autologous pericardium or expanded polytetrafluoroethylene was applied. Baseline, perioperative, and outcomes were analyzed.

***Results:*** No early mortality or perioperative complication occurred in Group A, while 2 patients died and 16 patients suffered from conduits failure due to conduits stenosis (11), stenosis plus regurgitation (n=3), and regurgitation alone (n=2) in Group B. Freedom from BJV grafts failure within 1, 3, 5, and 7 years was 98.0%, 88.2%, 83.6% and 83.6% in Group A, and 98.0%, 85.8%, 76.8% and 62.1% in Group B.

Endocarditis occurred in 9 patients in Group B, but not in Group A. Subsequent analysis showed that endocarditis is the only significant predictor of BJV grafts failure (odds ratio: 6.202, 95% confidence intervals 1.237~31.108).

***Conclusions:*** The novel simplified hand-sewn trileaflet valved conduits seems to be associated with lower incidences of perioperative complication, graft failure, and early-phase mortality, as compared with conventional BJV grafts.

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1:50 PM - 3:20 PM (Sat. Jul 8, 2017 1:50 PM - 3:20 PM ROOM 3)

## [II-JCKO3-05] Mid-term Outcomes of Survival and Quality of Life in Children with Complex Congenital Heart Diseases after atrioventricular valvuloplasty in China

○Li Xiang, Yiwei Liu, Yuan Huang, Hao Zhang (Center of Pediatric Cardiac Surgery,National Center for Cardiovascular Diseases and Fuwai Hospital, Chinese Academy of Medical Sciences, Peking Union Medical College, Beijing, China)

**Objectives**: To describe mid-term outcomes of survival and quality of life of children with complex congenital heart diseases (CCHD) after atrioventricular valvuloplasty.
**Methods**: From 2012 to 2015, 181 children with CCHD who underwent atrioventricular valvuloplasty were enrolled. Median age was 13.6 (7.4 - 34.2) months. The patients were divided into 4 groups. Group 1 received mitral valve repair and associated complicated cardiac anomalies repair (n=41), Group 2 received total endocardial cushion defect (TECD) correction (n=102), Group 3 received tricuspid valve repair for Ebstein anomaly (n=28) and Group 4 received atrioventricular valvuloplasty for single ventricle (SV) (n=10). The parents reported the household income and completed the pediatric Quality of Life Inventory (PedsQL), which including generic core scales and cardiac module, during the outpatient visit.
**Results**: The surgical mortalities were 2.4%, 2.9%, 0, and 20% in group 1 to 4, respectively ($p<0.05$). The median follow-up time was 32 months. During the follow-up, 6 patients died, and 2 in Group 4 .The lowest PedsQL scores and more cardiac symptom and cognitive problems were observed in Group 4. Meanwhile, the family with low household income had the significantly reduced number of outpatient visits.
**Conclusions**: The valvuloplasty for children with SV had the highest mortality and worst life quality. Although valvuloplasty with mitral valve, TECD and Ebstein anomaly had excellent surgical outcomes and acceptable life quality, the family socioeconomic status had negative effect on the closed follow-up.

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1:50 PM - 3:20 PM (Sat. Jul 8, 2017 1:50 PM - 3:20 PM ROOM 3)

## [II-JCKO3-06] A Novel Arterial Cannulation Technique for Selective Regional Cerebral Perfusion in Aortic Arch Surgery in Infants

○Hyungtae Kim[1], Si Chan Sung[1], Kwang Ho Choi[1], Hyoung Doo Lee[2], Geena Kim[2], Hoon Ko[2] （1.Department of Thoracic and Cardiovascular Surgery, Pusan National University Yangsan Hospital, Korea, 2.Department of Pediatrics, Pusan National University Yangsan Hospital, Korea）

**Background:** A selective regional cerebral perfusion (SRCP) has been widely adopted for brain protection in neonatal or infant aortic arch surgeries. We have developed a specially designed cannula tip that allows bidirectional blood flow and provides secure positioning of the polytetrafluoroethylene (PTFE) tube graft at the innominate artery.
**Methods:** Since January 2015, aortic arch reconstructions using a new arterial cannulation technique have been performed in 51 consecutive neonatal or infant patients. The median age and weight at the time of operation were 7 days (1-118 days) and 3.19 kg (1.87-5.84 kg), respectively. A longitudinal purse-string suture was placed at the innominate artery above the innominate vein, and the newly developed cannula tip connecting PTFE tube graft was introduced into the purse-string suture site after clamping of the innominate artery. The PTFE tube graft was then connected to the arterial cannula.
**Results:** There was no bleeding in the arterial cannulation site during the operation in all cases. Mean cardiopulmonary bypass time was 142.2±47.5 min, and mean SRCP time was 34.0±9.4 min. There was no patient who had an event of high pressure more than 200 mmHg at the operation. There was no cannulation site stenosis or aneurysmal change in postoperative CT angiography in all cases for a mean follow-up of 11.5±7.2 months. No neurologic dysfunction was noted after the operation.
**Conclusion:** Our arterial perfusion system employing a specially designed cannula tip enables performing SRCP through the innominate artery in an easier and safer manner.

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1:50 PM - 3:20 PM (Sat. Jul 8, 2017 1:50 PM - 3:20 PM ROOM 3)

# [II-JCKO3-07] Totally thoracoscopic without robotic assistance in cardiac surgery: the initial experience in Viet Nam and future perspectives

○Le Ngoc Thanh, Nguyen Cong Huu, Dang Quang Huy （Cardiovascular Center, E Hospital, Hanoi, Vietnam）

**Objectives:** To investigate the feasibility and safety of totally thoracoscopic without robotic assistance in cardiac surgery. **Patients and methods**: From 5/2016 to 3/2017: 45 conscutive patients have been operated (40 cases of ASD closure, 3 *Cor triatriatum*, 1partial AVSD repair, 1left atrial myxoma resection. Peripheral CPB was done via femoral artery, femoral veinous and internal jugular vein . CO2 was flowed into pericardial cavity. ASDs(40 patients) were closed with patch on beating heart, normothermia without aortic clamping. The other(5patients) with cardiac arrest used a trans-thoracic Chitwood aortic clamp, antegrade Custodiol HTK cardioplegic solution administration via a needle inserted into ascending aortic. **Results:** All the operations were successful. Neither patient required conversion to sternotomy nor thoracotomy. There didn' t have any hospital death or complications. Mean operation time and mean cardiopulmonary bypass time were 247.2 ± 44.6 (min) and 126.1±33.6 (min), respectively. These patients were extubated within the first 6 hours, the volume of blood drainage on the first day were less than 80 ml. Patients were able to return to normal activities 1 week postoperatively. **Conclusion:** Totally thoracoscopic without robotic assistance in cardiac surgery is safe and feasible alternative to the conventional full sternotomy approach in some simple types of heart

diseases. It has advantages of decreased postoperative pain, short recovery period, less surgical trauma and high cosmetic value with greater patient satisfaction.

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1:50 PM - 3:20 PM (Sat. Jul 8, 2017 1:50 PM - 3:20 PM ROOM 3)

# [II-JCKO3-08] A study on the optimal extra-cardiac conduit size by comparison of exercise capacity according to conduit size in patients with Fontan circulation.

○Sang-Yun Lee[1], Seong-Ho Kim[1], So-Ick Jang[1], Soo-Jin Park[1], Eun-Young Choi[1], Hye-Won Kwon[1],Ji-Seok Bang[1], Ja-Kyung Yoon[1], Chang-Ha Lee[2], Eun-Seok Choi[2], Sung-Kyu Cho[2], (1.Department of Pediatrics, Sejong General Hospital, Bucheon-si, Korea, 2.Department of Cardiovascular and Thoracic Surgery, Sejong General Hospital, Bucheon-si, Korea)

**Introduction**: Because Fontan circulation doesn' t have a subpulmonary ventricle, the preload is limited. In Fontan circulation with extra-cardiac conduit, the size of conduit could be an important factor in determining the preload. We compared exercise capacity with each conduit size and tried to search for optimal conduit size in Fontan circulation.

**Patients and Methods**: We reviewed the medical record of 289 patients with Fontan circulation. Patients who had other type Fontan circulation (Kawashima, atriopulmonary, lateral tunnel), SpO2 <85%, protein loosing enteropathy, chronotropic incompetence, and respiratory exchange ratio <1.0 were excluded. As a result, 96 patients were enrolled and classified according to conduit size. We compared with their exercise capacity and analyzed correlation between exercise capacity and conduit size per body surface area (BSA).

**Results:** Male was 53 and age was 17.8±5.5 years old. In cardiac catheterization, central venous pressure (CVP) was 12.4±2.5 mmHg and pulmonary vascular resistance (PVR) was 1.15±0.53 wu·㎡. In cardiopulmonary exercise test, predictive peak VO2 was 60.2±10.4% and VE/VCO2 was 35.9±7.2. In comparison of CVP, PVR, Qs, peak VO2, and VE/VCO2, each conduit groups did not have a difference. However, conduit size per BSA had a significant curved correlation with predictive peak VO2 and VE/VCO2.

**Conclusions:** Our results showed that patients with about 13 mm/㎡ conduit per BSA have the best exercise capacity. And, patients with larger than smaller sized conduit were found to be more attenuated in their ability to exercise.

JCK Oral

# JCK Oral 4 (II-JCKO4)

## Kawasaki Disease/General Cardiology 1

Chair:Masahiro Ishii(Department of Pediatrics, Kitasato University School of Medicine, Kanagawa, Japan)
Chair:Min Huang(Shanghai Children's Hospital, China)
Chair:Seong-Ho Kim(Department of Pediatrics, Sejong General Hospital, Korea)
Sat. Jul 8, 2017 3:30 PM - 4:20 PM ROOM 3 (Exhibition and Event Hall Room 3)

### [II-JCKO4-01] Severity assessment of coronary artery aneurysms in Kawasaki disease using the internal diameter Z-score

○Masaru Miura[1,2], Tohru Kobayashi[2], Naoya Fukushima[1,2], Taichi Kato[2], Shinya Shimoyama[2], Kenji Suda[2], Shigeto Fuse[2], Jun Maeda[2], Tsutomu Saji[2] （1.Department of Cardiology, Tokyo Metropolitan Children's Medical Center, Japan 2.The Z-Score Project 2nd Stage Study (ZSP2) Group, Japan）

3:30 PM - 4:20 PM

### [II-JCKO4-02] Identification of susceptibility genes associated with Kawasaki disease by targeted enrichment of genomic region sequencing technique

○Danying Zhu[1], Han Zhang[1], Sirui Song[1], Lijian Xie[1], Feng Qiu[2], Jing Yang[2],Tingting Xiao[1], Min Huang[1], （1.Department of Cardiology,Shanghai Children's Hospital Affiliated to Shanghai Jiaotong University,Shanghai,China, 2.Shanghai Center for Bioinformation Technology）

3:30 PM - 4:20 PM

### [II-JCKO4-03] Kawasaki shock syndrome complicated with macrophage activation syndrome

○Yao Lin, Lin Shi, Yanjun Deng, Yang Liu, Ping Lu （Department of Cardiology,Capital Institute of Pediatrics,Beijing,China）

3:30 PM - 4:20 PM

### [II-JCKO4-04] The change of serum inflammatory meditators in 40 children with Kawasaki disease

○Wang Yun[1], Wang Yang[2], Ji Xiao Dan[2], Zhou Nan[3] （1.Department of Pediatrics, Beijing New Century Women's and Children's Hospital, Beijing, China, 2.New Century Beijing Children’s Hospital, P.R. of China, 3.Beijing Children’s Hospital, P.R. of China）

3:30 PM - 4:20 PM

### [II-JCKO4-05] Evaluation of coronary vascular destruction by Optical Coherence Tomography and coronary artery diameter in acute phase of Kawasaki disease

○Ryuji Fukazawa, Yoshiaki Hashimoto, Makoto Watanabe, Kanae Tsuno, Koji Hashimoto, Miharu Akao, Mitsuhiro Kamisago, Yasuhiro Katsube （Department of Pediatrics, Nippon Medical School, Japan）

3:30 PM - 4:20 PM

3:30 PM - 4:20 PM (Sat. Jul 8, 2017 3:30 PM - 4:20 PM ROOM 3)

# [II-JCKO4-01] Severity assessment of coronary artery aneurysms in Kawasaki disease using the internal diameter Z-score

○Masaru Miura[1,2], Tohru Kobayashi[2], Naoya Fukushima[1,2], Taichi Kato[2], Shinya Shimoyama[2], Kenji Suda[2], Shigeto Fuse[2], Jun Maeda[2], Tsutomu Saji[2] (1.Department of Cardiology, Tokyo Metropolitan Children's Medical Center, Japan 2.The Z-Score Project 2nd Stage Study (ZSP2) Group, Japan)

**BACKGROUND** The Z-score of the internal diameter has recently been used to classify coronary artery aneurysms (CAA) in Kawasaki disease (KD), but its predictive value for coronary events (CE) is uncertain.
**OBJECTIVES** The aim of this study was to clarify the relationship of the Z-score to time-dependent CE occurrence in KD patients with CAA.
**METHODS** We surveyed 1,006 consecutive KD patients aged less than 19 years old, who received a coronary angiography between 1992 and 2011 in Japan. The time-dependent occurrence of CE including thrombosis, stenosis, obstruction, acute ischemic events, and coronary interventions was analyzed for small (Z-score <5.0), medium (≥ 5.0 to <10.0), and large (≥ 10.0 or actual internal diameter ≥ 8.0 mm) CAA.
**RESULTS** The 10-year event-free survival rate for CE was 100.0, 96.0, and 66.2% for the right coronary artery (RCA) (p <0.001), and 100.0, 95.7, and 65.2% for the left coronary artery (LCA) (p <0.001), for small, medium, and large CAA. Cox regression analysis revealed that large CAA (hazard ratio 6.49, 95% confidence interval 3.44 - 12.25) and male gender (3.10, 1.63 - 5.89) were independent risk factors for CE in the RCA. Similarly, large CAA (6.34, 3.19 -12.59) and male gender (2.40, 1.12 - 5.16) were independent risk factors for CE in the LCA.
**CONCLUSIONS** In KD patients, a large CAA based on the internal diameter Z-score and male gender were independent risk factors for CE. Male KD patients with a large CAA should be carefully observed to prevent CE.

3:30 PM - 4:20 PM (Sat. Jul 8, 2017 3:30 PM - 4:20 PM ROOM 3)

# [II-JCKO4-02] Identification of susceptibility genes associated with Kawasaki disease by targeted enrichment of genomic region sequencing technique

○Danying Zhu[1], Han Zhang[1], Sirui Song[1], Lijian Xie[1], Feng Qiu[2], Jing Yang[2],Tingting Xiao[1], Min Huang[1], (1.Department of Cardiology,Shanghai Children's Hospital Affiliated to Shanghai Jiaotong University,Shanghai,China, 2.Shanghai Center for Bioinformation Technology)

**Objective**
To discover susceptibility genes associated with Kawasaki disease（KD） and coronary artery lesion（CAL） through targeted enrichment of genomic region sequencing technique.
**Methods**
114 KD patients and 45 outpatients for health examination were recruited from Shanghai children's hospital between November 2015 and November 2016. Patients were dvided into two groups on the basis of echocardiography, one is KD with CAL and another is KD without CAL. 472 single nucleotide

polymorphisms associated with KD susceptibility genes and 512 genes in T cell receptor signaling pathway, toll-like receptor signaling pathway, Cytokine receptor interaction, TGF-beta signaling pathway were selected as targeted genes, and target exome capture sequencing chip were customized. Then use Illumina HiSeq X10 for high-throughput sequencing. The sequencing datas were used to find out susceptibility genes associated with KD and CAL.

**Results**

There are 26 susceptibility genes associated with KD and 21 with CAL. *RPS6KB, VAV1, ACVR2B* and *CXCL14* are significantly associated with KD. *CCL4*、*TNFRSF12A*、*IFIH1*and *IL26* are significantly genes of CAL formation. *CXCL14*(rs1046092) T allele(OR=11.455, 95%CI=1.531-85.736), *CXCL14*(rs2547) G allele(OR=11.070, 95%CI=1.477-82.972) increased the risk of KD. *CCL4*(rs1719144) G allele(OR=4.132, 95%CI=1.655-10.316), *CCL4*(rs1049807) A allele(OR=4.132, 95%CI=1.655-10.316), *CCL4* (rs1719152) T allele (OR=3.756, 95%CI=1.495-9.437) increased the risk of CAL (P <0.05).

**Conclusions**

Targeted sequencing technology can be used in children to evaluate the risk of KD and CAL.

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3:30 PM - 4:20 PM (Sat. Jul 8, 2017 3:30 PM - 4:20 PM ROOM 3)

## [II-JCKO4-03] Kawasaki shock syndrome complicated with macrophage activation syndrome

○Yao Lin, Lin Shi, Yanjun Deng, Yang Liu, Ping Lu （Department of Cardiology,Capital Institute of Pediatrics,Beijing,China）

We describe a case of Kawasaki shock syndrome (KDSS) in a 5-month infant who was complicated by macrophage activation syndrome (MAS). The boy had developed typical Kawasaki disease. After the first administration of intravenous immunoglobulin (IVIG) on the $6^{th}$ day at local hospital, he still had a persistent fever and elevated inflammatory markers 48 hours later without coronary artery dilation. He was admitted to our department on the $10^{th}$ day of illness and treated with IVIG (2 g/kg given as a single intravenous infusion) and aspirin (30–50 mg/kg/d). However, he still got a hypotension which showed a typical KDSS. Furthermore, blood cell (neutrophils, platelets and hemoglobin), fibrinogen and NK cell activity decreased, serum ferritin, cytokin and soluble CD25 concentration increased significantly, which showed signs of MAS. In addition, progressive coronary aneurysm formation was observed with the widest diameter of 9mm. On the basis of anti-shock therapy (including fluid resuscitation, human albumin intravenous infusion, and vasoactive agents), therapy for MAS was administrated immediately (methylprednisolone 4-6mg/kg/d tapering for 8 weeks and blood component transfusion). Clinical improvement was obtained 2 days later, and the patient was discharged 30 days later. Two months later, the boy was well developed with coronary aneurysm retraction to 7.3mm. KDSS and MAS are both severe and life-threatening complications of Kawasaki disease, therefore, early diagnosis and timely treatment is very important to save lives.

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3:30 PM - 4:20 PM (Sat. Jul 8, 2017 3:30 PM - 4:20 PM ROOM 3)

## [II-JCKO4-04] The change of serum inflammatory meditators in 40 children with Kawasaki disease

○Wang Yun[1], Wang Yang[2], Ji Xiao Dan[2], Zhou Nan[3] （1.Department of Pediatrics, Beijing New Century Women's and Children's Hospital, Beijing, China, 2.New Century Beijing Children's Hospital, P.R. of China, 3.Beijing Children's Hospital, P.R. of China）

**Objective** 40 KD children were retrospectively studied, we analyzed the clinical features and inflammatory meditators within different group,to explore further treatment and development of those KD children. **Methods** 40 KD children were divided into two groups, positive and negative history of allergic diseases, their clinical experiences and outcomes were recorded. 10 allergic rhinitis children and 10 fe- brile patients were also recruited in this study. **Results** 1) 25.0% (10/40) KD children have a history of allergic disease, the proportion of allergic dermatitis (30.0% vs. 9.2%) in the positive history group was significantly higher than those in negative history group during the convalescence phase($p<0.01$),The proportion of cardiology damage in the positive history group was also significantly higher than that the negative group at the acute phase ($p<0.05$). 2) The high elevation of IgE and CRP levels existed in KD acute phase, and there was a statistical difference in KD patients compared with allergic rhinitis and fe- brile controls, respectively ($p<0.01$). 3)TEC in the positive history group was not significantly higher than the negative group,$p>0.05$; But, the elevation of IgE levels continually existed in 5 KD children of after 3 month of IVIG therapy. **Conclusions** The proportion of allergic dermatitis in KD children with positive allergic diseases history is higher than in those with negative history; The levels of IgE were continually elevated in 5 KD children after IVIG. Further study should enrolled more KD children in order to explore the development of disease .

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3:30 PM - 4:20 PM (Sat. Jul 8, 2017 3:30 PM - 4:20 PM ROOM 3)

# [II-JCKO4-05] Evaluation of coronary vascular destruction by Optical Coherence Tomography and coronary artery diameter in acute phase of Kawasaki disease

○Ryuji Fukazawa, Yoshiaki Hashimoto, Makoto Watanabe, Kanae Tsuno, Koji Hashimoto, Miharu Akao, Mitsuhiro Kamisago, Yasuhiro Katsube （Department of Pediatrics, Nippon Medical School, Japan）

Background
Kawasaki Disease (KD) induces panvasculitis, and results in destruction of vascular layers. Destruction of inner elastic lamina cause infiltration of medial smooth muscle cell (SMC) into intima. And SMC, which causes transformation, leads to vascular remodeling accompanied by stenosis/occlusion of the coronary artery in remote stage. Optical Coherence Tomography (OCT) can identify destructive findings of the three-layer structure of coronary vessels. We examine how the disruption of vascular structure by OCT correlates with the diameter of coronary artery in acute phase.
Methods
OCT could be performed in chronic phase of nine KD cases. Their coronary diameter in acute stage was extracted from the acute phase echocardiogram findings, or coronary angiogram findings (CAG) executed within 6 months after onset of KD. The coronary artery was divided into nine segments of RCA: Seg. 1, 2, 3, 4, LCA: Seg. 5, 6, 7, 8, 11. In OCT, the presence or absence of vascular breakage was evaluated. Correlation between the coronary diameter and the presence or absence of circumferential 3-layer destruction of the coronary artery was examined.
Results

The relation of coronary diameter and the presence or absence of 3-layer destruction is, presence n=17: 4.8±1.5mm, absence n=12: 2.6±1.1mm (p=0.002), in echocardiography, and presence n=15: 4.9±1.7mm, absence n=27: 2.3±0.8mm (p<0.001), in CAG. Destructive findings of 3-layer structure increased both in echocardiogram and CAG when the coronary artery diameter exceeded 4 mm.

Conclusions

When the acute coronary artery diameter exceeds 4 mm, the risk of coronary artery structural destruction increases. In order to accurately predict coronary remodeling at the remote period, factors other than diameter, such as region of CAL or degree of inflammatory response in the acute phase, should also be considered.

JCK Oral

# JCK Oral 5 (II-JCKO5)

## Catheter Intervention

Chair:Satoshi Yazaki(Division of Pediatric Cardiology, Sakakibara Heart Institute, Tokyo, Japan)
Chair:Hong Gu(Department of Pediatric Cardiology, Beijing Anzhen Hospital, China)
Chair:Young-Hwue Kim(Department of Pediatric Cardiology, Congenital Heart Disease Center, Asan Medical Center, Korea)
Sat. Jul 8, 2017 4:20 PM - 5:10 PM ROOM 3 (Exhibition and Event Hall Room 3)

### [II-JCKO5-01] New self-expandable percutaneous pulmonary valve implantation using knitted nitinol-wire stent mounted with a tri-leaflet porcine pericardial valve

○Gi Beom Kim, Mi Kyung Song, Eun Jung Bae, Chung Il Noh （Department of Pediatrics, Seoul National University Children's Hospital, Seoul, South Korea）

4:20 PM - 5:10 PM

### [II-JCKO5-02] Stent of PDA as an alternative to Blalock -Taussig Shunt: single center experience

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

4:20 PM - 5:10 PM

### [II-JCKO5-03] Tricuspid valve annulus Z score before and after transcatheter closure of atrial septal defect

○ChunMin Fu[1], Ming-Tai Lin[2], Jou-Kou Wang[2] （1.Department of Pediatrics, National Taiwan University Hospital, Hsinchu Branch, Taiwan, 2.Pediatric Cardiology, Department of Pediatrics, National Taiwan University Children's Hospital, Taiwan）

4:20 PM - 5:10 PM

### [II-JCKO5-04] Hybrid pulmonary vein stenting in the patients with refractory to surgical pulmonary vein stenosis repair

○JaKyoung Yoon[1], Mi Kyung Song[2], Gi Beom Kim[2], Eun Jung Bae[2], Chung Il Noh[2], Jae Gun Kwak[3],Woong Han Kim[3], Jeong Ryul Lee[3], （1.Department of Pediatrics, Sejong General Hospital, Bucheon, South Korea, 2.Department of Pediatrics, Seoul National University Children’s Hospital, Seoul, South Korea, 3.Department of Thoracic and Cardiovascular Surgery, Seoul National University Children’s Hospital, Seoul, South Korea）

4:20 PM - 5:10 PM

### [II-JCKO5-05] A Hybrid Biventricular CRT for a 6-year-old patient with heart failure after interventional closure of ventricular septal defect

○Yiwei Chen, Fen Li, Wei Ji, Jinjin Wu, Lijun Fu, Jie Shen,Shumin Wang, （Department of Cardiology, Shanghai Children's Medical Center Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai, PR China）

4:20 PM - 5:10 PM

4:20 PM - 5:10 PM (Sat. Jul 8, 2017 4:20 PM - 5:10 PM ROOM 3)

## [II-JCKO5-01] New self-expandable percutaneous pulmonary valve implantation using knitted nitinol-wire stent mounted with a tri-leaflet porcine pericardial valve

○Gi Beom Kim, Mi Kyung Song, Eun Jung Bae, Chung Il Noh （Department of Pediatrics, Seoul National University Children's Hospital, Seoul, South Korea）

OBJECTIVE
To report first human cases of new self-expandable percutaneous pulmonary valve implantation (PPVI) using knitted nitinol-wire stent mounted with a tri-leaflet porcine pericardial valve.

METHODS
New valved-stent was made by knitted nitinol-wire backbone and tissue valve using porcine pericardium with multiple steps for tissue preservation including decellularization and alpha-galactosidase treatment. And, a feasibility study was done.

RESULTS
Ten patients underwent total correction of Tetralogy of Fallot previously and showed severe PR (mean PR fraction: 44.6%) and enlarged RV volume (mean indexed RV end-diastolic volume; 184.1 mL/$m^2$). Their median age at PPVI was 21.8 years old (range: 13-36). Five patients were implanted with 28 mm and 5 patients were implanted with 26 mm diameter valved-stent loaded in the 18 French delivery sheath. There were no significant peri-procedural complications in all patients. After procedure, there was no significant pulmonary stenosis or PR from echocardiography in all patients. All patients discharged 4 days after PPVI without any problem. Eight patient completed 6 months follow-up until now. Cardiac MRI showed that mean indexed RV end-diastolic volume decreased from 188.6 to 124.9 mL/$m^2$.

CONCLUSION
A feasibility study about first human implantation of new self-expandable percutaneous pulmonary valve using knitted nitinol wire mounted with a tri-leaflet porcine pericardial valve was completed and short-term follow-up data showed good result and there was no serious adverse effect associated with valve itself.

4:20 PM - 5:10 PM (Sat. Jul 8, 2017 4:20 PM - 5:10 PM ROOM 3)

## [II-JCKO5-02] Stent of PDA as an alternative to Blalock -Taussig Shunt: single center experience

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

BACKGROUND: assess the characteristics, indications,technique, safety, efficacy, short-term outcome, and complications of patent ductus arteriosus (PDA) stenting in duct-dependent congenital heart diseases as an alternative to surgical shunt.
METHODS: Between May 2012 and Aril 2013, 24 patients with duct-dependent congenital heart diseases

underwent PDA stenting as first palliative procedure.
RESULTS: Characteristics: mean age (days) is 56.8+ 76.4 (2 to 225). Mean weight (Kg) is 3.6+1.3 (2.1 to 7.7). Indications: pulmonary atresia with intact ventricular septum (5; 20.8%), pulmonary atresia with ventricular septum defect (13; 54.2%), pulmonary stenosis (4, 16.7%), D-TGA (1;4.2%) and Tetralogy of Fallot (1; 4.2%). Technique: in 20 patients (83.3%) stents were implanted by retrograde arterial approach and 4 patients (26.7%) by antegrade femoral venous approach. In 14 patients (54.2%) stent were implanted with coronary stent, 10 patients with renal stent. Results: PDA stenting was successful in 18 patients (91.6%) and failed in 2 patients (8.4%). Saturation increased from 59.3 (45-75) (before stenting) to 87.2 (56-99) (after stenting). For those with successful procedure, 15 patients (62.5%) with wide patent stent, 6 patients (25%) died due to decreased saturation. Short-term complications:three patients (12.6%) stent dislodged and migrated. There was no procedure-related mortality.
CONCLUSION: PDA stenting is an attractive alternative to surgical shunt in a majority of patients with duct-dependent circulation on account of post operative morbidity and complications of surgical shunt.

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4:20 PM - 5:10 PM (Sat. Jul 8, 2017 4:20 PM - 5:10 PM ROOM 3)

# [II-JCKO5-03] Tricuspid valve annulus Z score before and after transcatheter closure of atrial septal defect

○ChunMin Fu[1], Ming-Tai Lin[2], Jou-Kou Wang[2] (1.Department of Pediatrics, National Taiwan University Hospital, Hsinchu Branch, Taiwan, 2.Pediatric Cardiology, Department of Pediatrics, National Taiwan University Children's Hospital, Taiwan)

Introduction:
Trans-catheter closure of atrial septal defect (ASD) is a commonly used procedure with excellent outcome and minimal complication. Interatrial shunt via ASD produces right ventricle (RV) volume overload, and closure of ASD may stop the progression and normalize RV size. Tricuspid valve (TV) annulus may be regarded as surrogate of RV size. In this study, we aimed to investigate the change of TV annulus before and after closure of ASD.

Methods:
We obtained patient lists of ASD secundum type from 2011 to 2015 with age 1 to 13 years-old in National Taiwan University Children' s Hospital. We retrospectively collected the demographic data, echocardiography parameters, and hemodynamic parameters at catheterization. We calculated TV annulus Z score before and after ASD closure and examined the association between the pre-catheterization parameter and post-catheterization Z score change.

Results:
For this population, the female to male ratio was 2.5 to 1, the mean/median age at catheterization was 4.2 years-old. The mean/median Z score of tricuspid valve before catheterization was 1.25. Pre-catheterization proBNP and Qp/Qs were positively associated with tricuspid valve Z score. TV Z score decreased to stationary state within a few months.

Conclusion:
TV annulus may be used as index for RV volume overload and recovery from trans-catheter closure of

ASD. Further study for closure of ASD in older population may give us implication of long term effect of volume overload from ASD.

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4:20 PM - 5:10 PM (Sat. Jul 8, 2017 4:20 PM - 5:10 PM ROOM 3)

## [II-JCKO5-04] Hybrid pulmonary vein stenting in the patients with refractory to surgical pulmonary vein stenosis repair

○JaKyoung Yoon[1], Mi Kyung Song[2], Gi Beom Kim[2], Eun Jung Bae[2], Chung Il Noh[2], Jae Gun Kwak[3],Woong Han Kim[3], Jeong Ryul Lee[3], (1.Department of Pediatrics, Sejong General Hospital, Bucheon, South Korea, 2.Department of Pediatrics, Seoul National University Children's Hospital, Seoul, South Korea, 3.Department of Thoracic and Cardiovascular Surgery, Seoul National University Children's Hospital, Seoul, South Korea)

Back ground:
Pulmonary vein stenosis(PVS) is still frustrating disease, especially in the patient with multiple severe PVS. We performed hybrid pulmonary vein(PV) stent placement successfully in the 5 patients who underwent multiple surgery for severe PVS after TAPVR repair.
Cases and review:
5 patients were identified between 2013 and 2016, who underwent hybrid PV stenting for malignant PVS. We performed hybrid PV stent placement on one patient who had progressive multiple PVS and severe pulmonary hypertension despite recurrent surgical PV widening after cardiac type TAPVR repair. After hybrid stenting with coronary drug eluting stent (DES) and surgical pulmonary vein angioplasty, PVS was relieved and pulmonary hypertension was improved. Other 2 patients had severe multiple PVS after mixed type TAPVR repair. These patients underwent hybrid PV stenting with bare-metal stent (BMS) and PV ballooning. Last 2 patients who was diagnosed as functional single ventricle with pulmonary atresia and supra-cardiac type TAPVR developed progressive severe PVS after TAPVR repair. These patients also underwent hybrid stent placement. Hybrid PV stenting resulted in sufficient relief of PVS to permit clinical stabilization in all patients. All patients received aspirin and clopidogrel after the operation. 3 patients had undergone several elective further catheterizations for PV ballooning or large stent insertion after hybrid stenting procedure.
Conclusion:
Even though the prognosis of severe multiple PVS is very poor, hybrid PV stent placement could be a good palliation in this patient group.

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4:20 PM - 5:10 PM (Sat. Jul 8, 2017 4:20 PM - 5:10 PM ROOM 3)

## [II-JCKO5-05] A Hybrid Biventricular CRT for a 6-year-old patient with heart failure after interventional closure of ventricular septal defect

○Yiwei Chen, Fen Li, Wei Ji, Jinjin Wu, Lijun Fu, Jie Shen,Shumin Wang, (Department of Cardiology, Shanghai Children's Medical Center Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai, PR China)

Cardiac resynchronization therapy (CRT) is an effective treatment for adult heart failure patients with wide QRS duration (QRS≥150ms).
The positive response in adult HF prompted exploration of the use of CRT in pediatric HF patients. However, the effectiveness of CRT in the pediatric population is difficult to evaluate because of the complex anatomic substrates of congenital heart disease (CHD).
In this case, we introduce a hybrid CRT system for a 6y old girl with HF and CLBBB after an interventional closure of ventricular septal defect(VSD) 2 year before.
A 6y old girl , with the weight of 22kg, 2 years after an interventional occluding operation for perimemberous VSD was admitted into the hospital due to the progressive dyspnea and poor stamina. After a regular combined anti-HF treatment for at least 12 month, she showed a progressive decrease EF(EF=18.9%) in echocardiography and CLBBB(QRS=172ms) in ECG. .
The patient accepted CRT implantation using a hybrid way: RA and RV leads were placed through left subclavian vein and LV lead was placed epicardially, through a mini-thoracotomy and tunneled up to the prepectoral pocket by cardiac surgeon. One month follow up showed the QRS duration reduce to 142ms and EF rise to 27.8%.
Although more follow up data need to be collected, this case and the hybrid implantation of CRT may be helpful in pediatric and CHD patients.

JCK Oral

# JCK Oral 6 (II-JCKO6)

## Long-term Outcome/ Heart Failure/Arrhythmia

Chair:Yoshiki Mori(Department of Pediatric Cardiology, Seirei Hamamatsu General Hospital, Japan)
Chair:Zhiwei Zhang(Department of Pediatric Cardiology, Guangdong General Hospital, China)
Chair:Si Chan Sung(Department of Thoracic and Cardiovascular Surgery, Pusan National University Yangsan Children's Hospital , Korea)
Sat. Jul 8, 2017 5:10 PM - 6:00 PM ROOM 3 (Exhibition and Event Hall Room 3)

### [II-JCKO6-01] Assessment of limited athletic participation school age after operation for severe congenital heart disease based on cardiovascular disease screening

○Satoru Iwashima[1], Satoshi Ueda[2], Keisuke Sato[1], Yasuhiko Tanaka[3] （1.Deaprtment of Pediatrics Cardiology, Chutoen General Medical Center, Kakegawa city, Japan, 2.Subcommittee for Discussing School Heart Examination Results of the Shizuoka Prefecture Medical Association, Japan, 3.Department of Pediatric Cardiology, Shizuoka Children’s Hospital, Japan）

5:10 PM - 6:00 PM

### [II-JCKO6-04] The fate of aortic valve after doubly commiitted juxtaarterial ventricular septal defect repair

○Hanna Jung, Young Ok Lee, Joon Yong Cho （Department of Thoracic and Cardiovascular Surgery, Kyungpook National University Hospital Korea, republic of Korea）

5:10 PM - 6:00 PM

### [II-JCKO6-05] The outcome of junctional ectopic tachycardia following repair of congenital heart defects

○Yaping Mi, Bing Jia, Yonghao Gui （Cardiovascular Center, Children's Hospital, Fudan University, Shanghai, China）

5:10 PM - 6:00 PM

5:10 PM - 6:00 PM (Sat. Jul 8, 2017 5:10 PM - 6:00 PM ROOM 3)

## [II-JCKO6-01] Assessment of limited athletic participation school age after operation for severe congenital heart disease based on cardiovascular disease screening

○Satoru Iwashima[1], Satoshi Ueda[2], Keisuke Sato[1], Yasuhiko Tanaka[3] (1.Deaprtment of Pediatrics Cardiology, Chutoen General Medical Center, Kakegawa city, Japan, 2.Subcommittee for Discussing School Heart Examination Results of the Shizuoka Prefecture Medical Association, Japan, 3.Department of Pediatric Cardiology, Shizuoka Children's Hospital, Japan)

Background: Mortality in severe congenital heart disease (SCHD) has dramatically decreased, and only few reports evaluated the school life activities (SLAs) of children with SCHD.Purpose: This study aimed to evaluate the SLAs of students with SCHD.Subjects and Methods: Based on information from the school-based echocardiographic screening program at Shizuoka prefecture, we retrospectively reviewed the SLAs of 58 students, including 39 with heterotaxy syndrome (Hetero) and 19 with hypoplastic left heart syndrome (HLHS), who received total cavopulmonary connection (TCPC). The limited athletic (LA) and non-limited athletic (non-LA) criteria were management levels A to D and E, respectively. We compared the clinical outcomes related to hemodynamic factors between the LA and non-LA students.Results: Of the participants, 19 were LA and 39 were non-LA students. No significant differences in fetal diagnosis and HLHS incidence were found. TCPC procedures tended to be performed earlier after birth. The LA students had undergone an increasing number of palliation procedures before achieving TCPC, increasing brain natriuretic peptide (BNP) levels, and deceasing SaO2 after TCPC. Receiver-operating characteristic curve revealed that >34.3 pg/ml after TCPC predicted LA SLAs.Conclusion: High BNP level and low SaO2 after TCPC are appropriate predictive markers of LA SLAs. Consensus should be established regarding the safety of the practice, optimal training regimen, and appropriate rehabilitation program to achieve non-LA SLAs.

5:10 PM - 6:00 PM (Sat. Jul 8, 2017 5:10 PM - 6:00 PM ROOM 3)

## [II-JCKO6-04] The fate of aortic valve after doubly commiitted juxtaarterial ventricular septal defect repair

○Hanna Jung, Young Ok Lee, Joon Yong Cho (Department of Thoracic and Cardiovascular Surgery, Kyungpook National University Hospital Korea, republic of Korea)

**Background**

In patients with doubly commiitted juxtaarterial ventricular septal defect (DCJA VSD), postoperative fate of the aortic valve remains unclear. Postoperative progression of aortic regurgitation (AR) sometimes occur despite early operation before the development of AR. This review is to identify the incidence and predictors of late AR progression after VSD repair. And to find out the optimal timing for VSD repair with or without right coronary cusp (RCC) prolapse or AR.

**Methods**

From Jan 2002 to Dec 2013, the medical records of 90 consecutive patients who underwent DCJA VSD repair alone at our hospital were reviewed. Preoperative evaluation showed 43 patients (48%) had RCC prolapse and 22 (24%) had AR. Among 22 patients, 4 had mild AR and no patients had more than

moderate. Operative approach was through the pulmonary artery in all patients.

**Results**

Operative survival was 100%. The median follow up period after VSD repair was 3.5 years. Among 90 patients, 4 patients showed postoperative progressive AR. Of the 90 patients with long-term echo follow up, 1 had mild AR, 2 had moderate AR. AR was improved in 18 patients.

**Conclusion**

Among patients with DCJA VSD, the incidence of AR progression after VSD repair was much lower than expected in our study. Early operation may be the keys to prevent progressive AR. In the present study, most patients underwent VSD repair at the age of <24months when had no more than faint AR. Regrettably, having few patients with late AR progression after VSD repair, it was unable to analysis the risk factors of postoperative AR progression.

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5:10 PM - 6:00 PM (Sat. Jul 8, 2017 5:10 PM - 6:00 PM ROOM 3)

## [II-JCKO6-05] The outcome of junctional ectopic tachycardia following repair of congenital heart defects

○Yaping Mi, Bing Jia, Yonghao Gui (Cardiovascular Center, Children's Hospital, Fudan University, Shanghai, China)

Objective: To analyze the incidence and outcome of postoperative junctional ectopic tachycardia (JET) in children.Methods: We collected demographics and perioperative data in patients undergoing cardiac surgery from January 2015 to December 2016. All the patients with JET received the stepwise treatment beginning with surface cooling and continuous intravenous dexmedetomidine. The continuous intravenous amiodarone will be added if the heart rate was not controlled.Results: There were 26 JET cases (1.86%, 26/1395), including 16 cases of VSD, 6 TOF, 2 CoA+VSD, 1 Taussig - Bing+IAA and 1 TGA+VSD. The age was from 27 days to 8 months (median: 89.5d). The weight was from 3.8kg to 7.5kg (median: 5.7kg). The JET cases occurred most frequently in the infants younger than six months old (24/26, 92.31%), and with no occurrence in the children older than one year. No related death occurred in the JET cases. Four cases were controlled only under the treatment of cooling and continuous intravenous dexmedetomidine. Mean ventilation time increased from 18.5h to 75h amongst the cases without and with JET (P<0.05). Meanwhile, CICU stay increased from 2d to 7.5d when JET occurred (P<0.05). Conclusions: Postoperative JET is particularly frequent in young infants after congenital cardiac surgery and correlates with increased mechanical ventilation time and CICU stay. The strategy of postperative treatment will be beneficial. Aggressive treatment with cooling, dexmedetomidine with/without amiodarone is mandatory.

JCK Oral

# JCK Oral 7 (III-JCKO7)

## Kawasaki Disease/General Cardiology 2

Chair:Mamoru Ayusawa(Department of Pediatrics and Child Health Nihon University school of Medicine & Itabashi Hospital, Japan)
Chair:Lucy Youngmin Eun(Department of Pediatric Cardiology, Yonsei University, Seoul, Korea)
Sun. Jul 9, 2017 10:15 AM - 11:05 AM ROOM 3 (Exhibition and Event Hall Room 3)

### [III-JCKO7-01] Experience of recurrent Kawasaki disease complicated by giant aneurysms showing only two of the main symptoms

○Chika Nakamura[1], Yukiko Kawazu[1], Shinji Kaichi[2], Etsuko Tsuda[3] (1.Department of Pediatrics, Toyonaka Municipal Hospital, Toyonaka-City, Osaka, Japan, 2.Department of Pediatric Cardiology, Hyogo Prefectural Amagasaki General Medical Center, Hyogo, Japan, 3.Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan)

10:15 AM - 11:05 AM

### [III-JCKO7-02] Clinical analysis of hospitalized children with Kawasaki disease based on E - Science model, single center registry from 2009 to 2016

○Sirui Song, Danying Zhu, Min Huang, Lijian Xie, Tingting Xiao (Department of Cardiology,Shanghai Children's Hospital Affiliated to Shanghai Jiaotong University,Shanghai,China)

10:15 AM - 11:05 AM

### [III-JCKO7-03] Kawasaki Disease with Atypical Presentation Masquerading as Severe Infection: a 10-year retrospective review in a Tertiary Hospital in Hong Kong

○Chi Yu Dennis Au, Nai Chung Fong, Cheuk Man Ronald Fung, Wai Yau Daniel Mak, Wai Lim Yiu, Yu Ming Fu (Department of Pediatrics and Adolescent Medicine, Princess Margaret Hospital, Hong Kong SAR, China)

10:15 AM - 11:05 AM

### [III-JCKO7-04] Coronary Artery Bypass Grafting in Children with Severe Coronary artery lesions after Kawasaki Disease

○Wenbo Zhang, Ming Ye, Bing Jia, Fang Liu, Lin Wu (Cardiovascular Center, Children's Hospital of Fudan University, Shanghai , China)

10:15 AM - 11:05 AM

### [III-JCKO7-05] Risk factors and implications of progressive coronary dilatation in children with Kawasaki disease

○Ming-Tai Lin, Ming-Yu Liu, Chia-Hui Wu, Jou-Kou Wang, Mei-Hwan Wu (Department of Pediatrics, National Taiwan University Hospital, Taiwan)

10:15 AM - 11:05 AM

10:15 AM - 11:05 AM (Sun. Jul 9, 2017 10:15 AM - 11:05 AM ROOM 3)

## [III-JCKO7-01] Experience of recurrent Kawasaki disease complicated by giant aneurysms showing only two of the main symptoms

○Chika Nakamura[1], Yukiko Kawazu[1], Shinji Kaichi[2], Etsuko Tsuda[3] (1.Department of Pediatrics, Toyonaka Municipal Hospital, Toyonaka-City, Osaka, Japan, 2.Department of Pediatric Cardiology, Hyogo Prefectural Amagasaki General Medical Center, Hyogo, Japan, 3.Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan)

We report a recurrent case of Kawasaki disease (KD) complicated by giant aneurysms that developed only fever and slight conjunctival hyperemia.
At 2 years old, the patient had complete KD and received intravenous immunoglobulin therapy. He was discharged on day 11 of the illness. 7 months later, he had developed a cough and fever. He was subsequently referred to us because of high inflammatory reaction. Upon admission, he had only 2 major symptoms of KD, fever and slight conjunctival hyperemia. He had a cough and a chest X-ray showed consolidation, so we suspected bacterial pneumoniae and started antibiotic therapy. This failed to bring his temperature down, so we tried another antibiotic drug, but that too was ineffective. Slight conjunctival hyperemia disappeared on the next day of admission, and other symptoms of KD hadn't appeared. On day 7 of the illness, ultrasound sonography showed bilateral coronary artery aneurysms like a string of beads, therefore we diagnosed him as incomplete KD, and administered intravenous immunoglobulin, cyclosporine A and aspirin. Next morning, the fever went down but recurred during the night. As we were afraid that the coronary artery aneurysms would expand further, he was transferred to another hospital where he received plasma exchange.
It is reported that patients with recurrent KD are at risk of coronary artery lesions, and tend to be atypical. This case developed only 2 major symptoms, we had difficulty making a correct diagnosis. When encountering children having fever with a history of KD, we should consider recurrence of KD.

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## [III-JCKO7-02] Clinical analysis of hospitalized children with Kawasaki disease based on E - Science model, single center registry from 2009 to 2016

○Sirui Song, Danying Zhu, Min Huang, Lijian Xie, Tingting Xiao (Department of Cardiology,Shanghai Children's Hospital Affiliated to Shanghai Jiaotong University,Shanghai,China)

**Objective** Based on the E-Science environment, the data of Kawasaki disease (KD) patients in Shanghai Children's Hospital from 2009 to 2016 were analyzed in order to provide the basis for clinical diagnosis and treatment. The study aimed to investigate the clinical characteristics and risk factors of coronary artery lesion (CAL) in KD patients with the registry database .
**Method** The children with KD who were hospitalized in Shanghai Children's Hospital,selected according to the American Heart Association (AHA) diagnostic guidelines ,the incidence of CAL was determined according to the results of cardiac ultrasonography(UCG), all clinical indexes had been tagged and captured by electronic data capture (EDC) system. Statistical analyses were conducted by the Doctor

Research Information Management System(DRIMS). Clinical indexes were performed to analyze the risk factors and incidence trend of CAL.

**Results** A total of 1157 cases were hospitalized mostly in spring and summer. The incidence of IVIG non-response was 13.1% . UCG found 22.92% cases of CAL, including 2.05% of coronary aneurysm. The trend of CAL incidence was declining by year. C-reactive protein (CRP) level, serum sodium level, and ALT > 40U/L or AST >40U/L were the influencing factors of CAL.

**Conclusion** Our findings highlight the frame of data management and analysis.The E-Science environment shows a good effect on the large-scale epidemiological investigation. There are a certain epidemic characteristics of Kawasaki disease in our single center.

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# [III-JCKO7-03] Kawasaki Disease with Atypical Presentation Masquerading as Severe Infection: a 10-year retrospective review in a Tertiary Hospital in Hong Kong

○Chi Yu Dennis Au, Nai Chung Fong, Cheuk Man Ronald Fung, Wai Yau Daniel Mak, Wai Lim Yiu, Yu Ming Fu (Department of Pediatrics and Adolescent Medicine, Princess Margaret Hospital, Hong Kong SAR, China)

**Objective**

Kawasaki disease (KD) can masquerade as severe infection: KD shock syndrome (KDSS) mimicking toxic shock; and retropharyngeal edema (RPE) mimicking neck abscess. They may delay diagnosis thus timely treatment.

**Method**

A 10-year (2007-2016) review was done for any KDSS / RPE.

**Result**

120 KD cases were recruited. M:F was 1.2:1, age 3.5 months-12 years old.

7 had RPE mimicking abscess (5.8%), presented as fever with neck complaints: swelling, pain or refusal to rotation/extension. None fulfilled diagnostic criteria of KD initially, some even with absent KD signs. They responded poorly to antibiotics. Neck X-ray showed thickened retropharyngeal space suspicious of abscess. Computer tomography (CT) showed retropharyngeal fluid with no definite rim-enhancement. One case in doubt of early abscess underwent fine needle aspiration yielding 1.5ml necrotic material, sterile for bacterial culture. KD signs emerged as clinical course progressed, and they responded well to immunoglobulin (IVIG) and aspirin.

3 presented as KDSS mimicking toxic shock (2.5%) with systolic hypotension needing inotropes and had ventricular dysfunction, mitral regurgitation on echocardiogram. Two had coronary ectasia on presentation. All were IVIG resistant which were well predicted by Egami score. Two required pulse methylprednisolone and remaining one responded to 2$^{nd}$ dose IVIG.

**Conclusion**

KD can present atypically leading to diagnostic confusion mimicking severe infections. High vigilance is

needed for early diagnosis and timely treatment to minimize unnecessary operations and complications.

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## [III-JCKO7-04] Coronary Artery Bypass Grafting in Children with Severe Coronary artery lesions after Kawasaki Disease

○Wenbo Zhang, Ming Ye, Bing Jia, Fang Liu, Lin Wu （Cardiovascular Center, Children's Hospital of Fudan University, Shanghai , China）

**Objectives** Severe coronary artery lesions (CALs) in affected children after Kawasaki disease (KD) usually need further assessment and coronary artery bypass grafting(CABG). CABG is the most effective procedure for this consequence and essential in preventing premature death and improving the quality of life for children. The aim of this study was to evaluate the efficacy of CABG for the treatment of severe coronary artery lesions in children after KD.
**Methods** The subject of the study was 9 patients with giant coronary aneurysms after KD who underwent CABG between Aug 2005 and July 2016, including 7 boys and 2 girls, aged from 2y~14y.Body weight at operation ranged from 12.5to 51 kg, with a media weight of 26kg. CALs were found by echocardiography (ECHO) and coronary artery angiography, all the 9 patients underwent CABG with left radial artery or/and left posterior descending artery.
**Results** There was only one hospital death. The remaining 8 patients received various combined medications. Dual antiplatelet medication therapy were used in all the 8 patients. There was no late deaths during the follow-up period. All patients can live a normal life and 6 are permitted most physical activities including long-distant swimming and running while other 2 are prohibited from strenuous exercise.
**Conclusion** CABG should be considered when myocardial ischemia was detected, and the patency rate is acceptable during the follow-up period.

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## [III-JCKO7-05] Risk factors and implications of progressive coronary dilatation in children with Kawasaki disease

○Ming-Tai Lin, Ming-Yu Liu, Chia-Hui Wu, Jou-Kou Wang, Mei-Hwan Wu （Department of Pediatrics, National Taiwan University Hospital, Taiwan）

**BACKGROUND**
Kawasaki disease (KD) is an acute systemic vasculitis in childhood. The risk factors and occurrence of progressive coronary dilatation in KD patients have been insufficiently explored.
**METHODS**
We retrospectively enrolled KD patients during 2009–2013. Echocardiography was performed during the acute KD phase and at 3–4 weeks, 6–8 weeks, 6 months, and 12 months after KD onset. Progressive coronary dilatation was defined as the progressive enlargement of coronary arteries on three

consecutive echocardiograms. Logistic regression analysis was conducted to evaluate the potential risk factors for coronary aneurysms and progressive coronary dilatation.

## RESULTS

Of a total of 169 patients with KD, 31 (18.3%) had maximal coronary Z-scores of ≥+2.5 during the acute KD phase, 16 had coronary aneurysms at 1 month after KD onset, and 5 (3.0%) satisfied the definition of progressive coronary dilatation. Multivariate logistic regression analysis revealed that an initial maximal coronary Z-score of ≥+2.5 [$P = 0.02$] and hypoalbuminemia ($P = 0.03$) were independent risk factors for coronary aneurysms and for progressive coronary dilatation.

## CONCLUSIONS

In the present study, 3% (5/169) of patients with KD had progressive coronary dilatation, which was associated with persistent coronary aneurysms at 1 year after KD onset. Initial coronary dilatation and hypoalbuminemia were independently associated with the occurrence of progressive coronary dilatation. Therefore, such patients may require intensive cardiac monitoring and adjuvant therapies apart from immunoglobulin therapies.

JCK Oral

# JCK Oral 8 (III-JCKO8)

## Cardiovascular Imaging

Chair:Keisuke Satou(Department of Cardiology Shizuoka Children Hospital, Japan)
Chair:Seong-Ho Kim(Department of Pediatrics, Sejong General Hospital, Korea)
Sun. Jul 9, 2017 11:05 AM - 11:55 AM ROOM 3 (Exhibition and Event Hall Room 3)

### [III-JCKO8-01] Morphological and clinical spectrum of infantile scimitar syndrome

○Lin Wu, Fang Liu （Department of Cardiology, Children's Hospital of Fudan University, Shanghai-City, China）
11:05 AM - 11:55 AM

### [III-JCKO8-02] New fusion imaging - VesselNavigator in CHD interventions

○Seong Ho Kim, Su Jin Park, Hye Won Kwon, Sang Yun Lee, Ji Seok Bang, Eun Young Choi, So Ick Jang, Ja Kyung Yun （Department of Pediatrics, Sejong General Hospital, Korea）
11:05 AM - 11:55 AM

### [III-JCKO8-03] Cardiac Mechanics in Children post Percutaneous Transcatheter Closure of Perimembranous Ventricular Septal Defect

○Lijian Xie, Xunwei Jiang, Yun Li, Min Huang, Tingting Xiao （Department of Cardiovascular, Shanghai Children's Hospital, Shanghai Jiaotong University, Shanghai, China）
11:05 AM - 11:55 AM

### [III-JCKO8-05] Evaluation of tricuspid valve displacement in repaired tetralogy of Fallot using feature tracking magnetic resonance program

○Akio Inage[1], Naokazu Mizuno[2] （1.Division of Pediatric Cardiology, Sakakibara Heart Institute, Japan, 2.Division of Radiology, Sakakibara Heart Institute, Japan）
11:05 AM - 11:55 AM

11:05 AM - 11:55 AM (Sun. Jul 9, 2017 11:05 AM - 11:55 AM ROOM 3)

## [III-JCKO8-01] Morphological and clinical spectrum of infantile scimitar syndrome

○Lin Wu, Fang Liu (Department of Cardiology, Children's Hospital of Fudan University, Shanghai-City, China)

**Objective** Scimitar syndrome is a rare constellation of cardio-pulmonary anomalies. We describe the morphological and clinical spectrum in a retrospective case series of infantile scimitar syndrome. **Methods** A total of 7 patients with infantile scimitar syndrome were identified in our hospital from 2009 to 2016. **Results** The mean age at diagnosis was 4.7±2.6 months with mean body weight of 5.2±1.0kg. All patients presented with symptoms, including respiratory distress (n=5), heart failure (n=4), recurrent pneumonia (n=3) and cyanosis (n=2). There were 3 patients associated with ASD, 2 with TOF and 1 with ASD and PDA. Anomalous right pulmonary scimitar vein provided drainage for the entire right lung in 5 patients, and only the lower segments in 2 patients, including drainage site to superior vena cava in 1 patient, to the hepatic vein with obstruction in 1 and to inferior vena cava in 5. All the patients underwent cardiac catheterization. Severe and moderate pulmonary arterial hypertension was demonstrated in 2 and 3 patients, respectively. Transcatheter collateral embolization was performed in 4 patients. Surgical repair of intra-cardiac lesion with and without anomalous vein correction was carried out in 2 and 3 patients, respectively. Mean follow-up time was 12.5±10.5 months. There were 2 hospital deaths and no late death. **Conclusions** The infantile form of scimitar syndrome is a very severe form of disease, usually associated with heterogeneous morphological variations. A careful anatomic study is mandatory for prompt treatment.

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## [III-JCKO8-02] New fusion imaging - VesselNavigator in CHD interventions

○Seong Ho Kim, Su Jin Park, Hye Won Kwon, Sang Yun Lee, Ji Seok Bang, Eun Young Choi, So Ick Jang, Ja Kyung Yun (Department of Pediatrics, Sejong General Hospital, Korea)

**Objectives**: VesselNavigator is a new fusion imaging modality which overlays 3D CT image onto a live fluoroscopy image. It provides an intuitive and continuous 3D roadmap by rotating overlaid CT imaging at any directions, and allows for both advanced diagnostic and interventional cardiac catheterization procedures in patients with congenital heart disease(CHD). Recently, in our institute it was available first in Asia. We would like to share our initial experiences with the audiences.
**Methods**: Between May 2016 and March 2017, VesselNavigator has been used in 14 patients with postoperative CHD; tetralogy of Fallot or pulmonary atresia in 7, transposition of great artery in 3, coarctation of aorta in 2 and others in 2. Mean age was 13.3(0.5 – 28)years, and mean body weight was 36.3(7 – 68)kg. It helped to access a target vessel easily, and to find out the best angiocamera angle for diagnosis and interventional procedure without additional angiography.
**Results**: VesselNavigator was used for pulmonary arterial interventions in 8(stent in 6, balloon angioplasty in 2), MAPCA occlusion in 1 and diagnosis in 5 patients. Mean fluoroscopic time and procedure time were 21(11 - 38) and 63(30 - 97) min., respectively.
**Conclusions**: VesselNavigator is a promising modality for CHD diagnosis and treatment. Using

VesselNavigator as a 3D roadmap without additional 3D rotational angiography, we can reduce fluoroscopic and procedural time, contrast amount and radiation exposure.

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11:05 AM - 11:55 AM (Sun. Jul 9, 2017 11:05 AM - 11:55 AM ROOM 3)

## [III-JCKO8-03] Cardiac Mechanics in Children post Percutaneous Transcatheter Closure of Perimembranous Ventricular Septal Defect

○Lijian Xie, Xunwei Jiang, Yun Li, Min Huang, Tingting Xiao （Department of Cardiovascular, Shanghai Children's Hospital, Shanghai Jiaotong University, Shanghai, China）

Background Percutaneous transcatheter closure of perimembranous ventricular septal defect (pmVSD) with occluder has been most widely used in China.In this study, we aimed to analyze ventricle performance post percutaneous transcatheter closure of pmVSD.
Methods 40 pediatric patients post percutaneous pmVSD closure and 40 healthy children were recruited. All subjects were studied with conventional and tissue Doppler echocardiography. Strain and strain rate of LV and RV were assessed by 2D-STE.
Results 40 pmVSD pediatric patientsand 40 healthy controlswere studied. Mean diameter of pmVSD was 3.82±0.59 mm, mean diameter of pmVSD occluder was 6.3±1.0mm, and mean time after percutaneous pmVSD closure was 3.22±0.78years. No significant differences were observed in LV eject fraction, RV Tei index between pmVSD closure and control. More tricuspid regurgitation was observed in pmVSD closure subjects by measuring the ratio of tricuspid regurgitation jet area and right atrial area. Interventricular septal tissue Doppler image showed less early diastolic, more late diastolic velocity and less e/a ratio in pmVSD closure subjects. No significant difference in global longitudinal and circumferential strain and strain rate between pmVSD closure and control.For pmVSD closure cohort, the diameter of pmVSD occluder correlated negatively with LV longitudinal strain rate and circumferential strain. Furthermore, TRJA/RAA correlated positively with diameter of pmVSD and occluder.
Conclusions It appears that percutaneous closure of pmVSD is safe and effective in selected pediatric patients.

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11:05 AM - 11:55 AM (Sun. Jul 9, 2017 11:05 AM - 11:55 AM ROOM 3)

## [III-JCKO8-05] Evaluation of tricuspid valve displacement in repaired tetralogy of Fallot using feature tracking magnetic resonance program

○Akio Inage[1], Naokazu Mizuno[2] （1.Division of Pediatric Cardiology, Sakakibara Heart Institute, Japan, 2.Division of Radiology, Sakakibara Heart Institute, Japan）

**Introduction:** Novel cardiac MRI (CMR) software may provide a simple method to assess right ventricular (RV) myocardial deformation in tetralogy of Fallot (TOF). The objective of this study was to determine correlations between tricuspid valve (TV) displacement and RV function in patients with repaired TOF using CMR.

**Methods:** We retrospectively analyzed 25 CMR studies in repaired TOF (24+/-8 years). Peak RV longitudinal strain, systolic strain rate (SR) and end-diastolic SR (EDSR) were measured from 4-chambers using cine-based feature tracking MR program. TV displacement was measured at end-systole as the shortest distance between both anterior and septal leaflet hinge points relative to the RV apex. Basal anterior and septal displacement velocities in systole and early diastole were computed. We investigate into correlation TV displacement parameters with RV functional parameters.
**Results:** Increased anterior and septal distances as decreased shortening correlated positively with RV end-diastolic volume and end-systolic volume, negatively with ejection fraction and longitudinal strain. Decreased basal anterior displacement velocities as greater shortening were associated with improved longitudinal strain, SR and EDSR; however, there were no correlations between basal septal displacement velocities and RV functional parameters.
**Conclusion:** Decreased TV shortening in systole is associated with larger RV volumes and decreased RV function. Greater anterior displacement velocities in systole and early diastole are associated with improved RV contractility and early filling rate.