

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 Nakamura¹, Yukiko Kawazu¹, Shinji Kaichi², Etsuko Tsuda³ (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)

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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)

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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)

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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 Inage¹, Naokazu Mizuno² (1.Division of Pediatric Cardiology, Sakakibara Heart Institute, Japan, 2.Division of Radiology, Sakakibara Heart Institute, Japan)

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

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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.

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

[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 2nd 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.

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

[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 $\geq +2.5$ 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 $\geq +2.5$ [$P = 0.02$] 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)

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[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)

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

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

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

○Akio Inage¹, Naokazu Mizuno² (1.Division of Pediatric Cardiology, Sakakibara Heart Institute, Japan, 2.Division of Radiology, Sakakibara Heart Institute, Japan)

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[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 ± 1.0 kg. 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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[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 patients and 40 healthy controls were studied. Mean diameter of pmVSD was 3.82 ± 0.59 mm, mean diameter of pmVSD occluder was 6.3 ± 1.0 mm, and mean time after percutaneous pmVSD closure was 3.22 ± 0.78 years. No significant differences were observed in LV ejection 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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[III-JCKO8-05] Evaluation of tricuspid valve displacement in repaired tetralogy of Fallot using feature tracking magnetic resonance program

○Akio Inage¹, Naokazu Mizuno² (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.