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CLINICAL STUDY OF 104 CASES WITH KAWASAKI DISEASE

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Objective To acquire the situation of pathogeresis, diagnosisand treatment of Kawasaki disease in Shannxi Province. **Methods** we had a retrospective clinical study on children with KD in six hospital in Xi'an between 1998 and 1999. This is a consecutive study based on the epidemic study on KD of Sino-Japanese between 1993 and 1997. The evaluation include age, sex, season, clinical symptoms and signs, the counting of thrombocyte, ESR, cardiac enzyme series. With the dose of IVGG, all the patients were designed into four groups: A 300~400mg/kg/d; B 400~800mg/kg/d; C 1000mg/kg/d ; D 2000mg/kg/d, together with aspirin and anticoagulant. **Results** Sex: male 73; female 31. Age: 1 year 24.03% ; 1~3 years 50.4%; 3~7 years 22.11%; 7~14 years 2.88%. 80.76% patients had fever persisting for fine days or more ; 82.69%, changes in the extremities ; 74.03%, polymorphous exanthema; 78.84%, changes in the lips and oral cavity ; 74.63%, nonpurulent cervical lymphalenopathy. There were 21 patients to be diagnosed as acute upper respiratory infection (AURI) by impression; 14, herpangina; 5, cervical lymphadenopathy; 2, lymphomas; 1, thrombocythemia. **Conclusion** According to recent study, the cardiovascular damage of KD was gradually increased, especially in coronal artery and became the main cause of acquired heart disease in Chinese children with KD. IVGG has been an effective therapy for KD in recent years, using in 400mg/kg/d for 4~5days was superior to 100~200mg/kg/d in China.

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NATURAL HISTORY OF CORONARY ARTERY ECTASIA IN CHILDREN WITH KAWASAKI DISEASE

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Coronary artery ectasia is the most common coronary artery lesion associated with Kawasaki disease (KD), and is felt to regress with no long-term sequelae. We sought to determine the natural history of ectasia and associated factors. From 1990-2001, 143 patients (69% male) were identified with sufficient echocardiographic documentation of ectasia. The median age at KD was 2.7 years (range, 2.3 months to 15 years), with 73% receiving IVIG. Aneurysms were also present in 31% (11% with giant aneurysms). Serial measurements of coronary artery dimensions of non-aneurysmal proximal segments were converted to Z scores based on body surface area (BSA). Patients had a median of 4 echocardiograms each (range, 2 to 15) over a median interval of 1.6 years (range up to 9.8 years). Mixed linear regression was used to determine trends and factors associated with the serial Z scores for the right (zRCA), left main (zLMCA) and left anterior descending (zLAD) coronary arteries. Initial mean (SD) zRCA was 0.60 (1.07), zLMCA 0.99 (1.28) and zLAD 0.87 (1.40). There were no significant trends in Z scores over time, indicating a lack of regression or progression when measurements were normalized for BSA. Further, no trend was noted when analysis was limited to arteries with initial Z scores above 2. Higher zRCA was independently related to no IVIG treatment ($p=0.08$), higher initial zRCA ($p=0.001$), lower initial zLMCA ($p=0.001$), and higher zLAD ($p=0.001$). Higher zLMCA was independently related to male gender ($p=0.01$), presence of any aneurysms ($p=0.01$), higher initial zLMCA ($p=0.03$), lower initial zRCA ($p=0.07$) and higher zLAD ($p=0.001$). Higher zLAD was independently related to higher initial zLAD ($=0.001$), higher initial zLMCA ($p=0.03$) and higher zRCA ($p=0.001$). In conclusion non-aneurysmal coronary artery segments appear to grow proportionately, and subsequent dimensions are related to initial dimensions and dimensions of other segments.

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OVERALL OUTCOME OF KAWASAKI DISEASE

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To assess the overall outcome of Kawasaki disease, patients with a discharge diagnosis of Kawasaki disease at Korea University Medical Center from 1999 to 2001 were retrospectively evaluated. A total of 99 patients were diagnosed. The American Heart Association (AHA) criteria were met in 65 patients (66%) and 63 of them received intravenous immune globulin (IVIG, 2g/kg). Fifty patients responded afebrile within 5 days (IVIG-responsive) and 13 patients did not (IVIG-non-responsive). Within 2 months, 2 patients in the IVIG-responsive group developed coronary abnormalities compared to 3 patients in the IVIG-non-responsive group (4% vs 23%). None of whom met AHA criteria and did not received IVIG were febrile more than 5 days nor developed coronary abnormalities. In 34 patients who did not meet complete AHA criteria, 28 patients received IVIG and 25 patients were IVIG-responsive. Coronary abnormalities were developed in 4 in the IVIG-responsive group compared to none in the IVIG-non-responsive group (16% vs 0%). All 6 patients who did not meet complete AHA criteria and did not received IVIG were febrile more than 5 days and 2 of them developed coronary abnormalities. Taken together, coronary abnormalities were developed in 9 patients of 91 who were treated with IVIG compared to 2 patients of 8 who were not treated with IVIG (10% vs 25%). In conclusion coronary abnormalities were developed in 11 of 99 patients with Kawasaki disease (11%).

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A CLINICAL STUDY OF ATYPICAL KAWASAKI SYNDROME: A COMPARISON WITH TYPICAL KAWASAKI SYNDROME

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The diagnosis of Kawasaki syndrome (KS) is based on the clinical features. Not all KS patients fulfill the classic diagnostic criteria and diagnosis is often based on the finding of coronary aneurysm by echocardiography. Children with KS manifested by fever and fewer than four other criteria, so called atypical KS, are known to be at high risk for coronary aneurysms. This study was performed to evaluate clinical and laboratory features including coronary artery involvement in atypical KS in comparison with typical KS. Data were reviewed retrospectively in 72 patients with KS. According to the diagnostic criteria, these patients were divided into 30 of atypical KS and 42 of typical KS. The mean duration of fever was 5.8±·4.3 days in atypical KS(6.7±4.8 days in typical KS, p0.05). The incidence of each diagnostic criteria in atypical KS were as follows: conjunctival injection 80%, changes of oral mucosa 73.3%, rash 43.3%, changes of hands and feet 36.6%, and cervical lymphadenopathy 23.3%. Erythema at the site of BCG was observed in 7 of 13 patients with atypical KS who were less than 25 months old. There was no significant difference in the incidence of other clinical features such as aseptic meningitis, gallbladder hydrops, arthralgia and arthritis. Echocardiographic abnormalities were observed in 20% of atypical KS (aneurysm 6.7% and ectasia 13.3%) and 47.7% of typical KS (aneurysm 14.3% and ectasia 33.4%) during the subacute phase. Follow-up echocardiographic studies during convalescent phase revealed regression of coronary artery abnormalities: 7.7% of atypical KS (aneurysm 0%, ectasia 7.7%) and 14.7% of typical KS (aneurysm 2.9% and ectasia 11.8%). Statistical analysis determined no significant difference in the incidence of coronary artery involvement between atypical and typical KS. In conclusion, atypical KS emphasize the index of suspicion of KS with respect to the coronary artery involvement.

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ATYPICAL KAWASAKI DISEASE- EXPERIENCE IN HONG KONG

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Introduction: Infants with atypical Kawasaki Disease always posed a diagnostic difficulty to cardiologist and literature has suggested that infants with Kawasaki Disease had a higher complication rate. A retrospective review of atypical Kawasaki disease from July 1994 to June 2000 was carried out to study the incidence and coronary complication rate among patients with atypical Kawasaki Disease. Methods: The hospital records of patients with Kawasaki diseases diagnosed between July 1994 to June 2000 were reviewed by members of Hong Kong Kawasaki Disease Study Group. Patients were identified as atypical Kawasaki disease if they have less than five of the diagnostic criteria after having excluded other possible causes. Results: A total of 72 cases of atypical Kawasaki disease were diagnosed from July 1994 to June 2000. Forty-five are males. Twenty-seven were females. M: F ratio 1.7: 1. Age of onset range from 0.2yr to 12.8yrs (mean 2.1yr; median 2.2yr). 39 of them are infants (54%; $p< 0.0001$). 56 (79%) received IVIG therapy ($p =0.01$). Coronary artery aneurysm developed in 25 cases (34.7%) of atypical Kawasaki Disease ($p<0.0001$). 15 (60%) of them were infants. 20 of the atypical cases with coronary aneurysm received IVIG therapy. Conclusion: We conclude that patients with atypical Kawasaki disease had a higher rate of coronary complication. Infants were associated with a higher rate of atypical presentation and coronary complication. Male sex and use of IVIG contribute no statistical significance in the development of coronary complication in atypical Kawasaki disease. However, patients with atypical Kawasaki disease had a significant lower incidence of receiving IVIG therapy when compared with typical Kawasaki presentation. This may be due to delay in diagnosis and treatment. A high index of suspicion and clinical assessment by cardiologist is recommended in the diagnosis and treatment of atypical Kawasaki disease especially among infants.

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CORONARY NORMOGRAMS AND CORONARY-AORTIC RATIO FOR DETERMINATION OF CORONARY DILATATION

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Echocardiographic measurements of the internal diameter of the left (LCA) and right (RCA) main coronary arteries were performed on 390 children (215 male, 175 female) with normal cardiac structures, who were within the ages of 2 months to 8 years. They have normal clinical examination and complete echocardiographic examination prior to enrollment for the study. The maximal internal diameters within the cardiac cycles were measured at a distance of 5 to 10 mm from the origins of the LCA and RCA.The racial composition of the study population comprises 73.3% Chinese, 17.4% Malay, 6.2% Indian and 3.1% others. Their mean age was 34 months (range 2 to 106 months). The mean weight and height were 13.0 ± 6.0 kg and 88.6 ± 20.9 cm, respectively. After controlling for age, height, weight and body surface area (BSA), the LCA and RCA diameters were larger in boys (mean difference 0.1mm, $p < 0.005$). The diameter of the LCA and RCA correlated linearly with age, height, weight and BSA (Pearson $R > 0.8$, $p < 0.005$). The regression equations, prediction graphs and z-score graphs are given. For the same patient, the LCA is generally larger than the RCA (mean difference 0.30 ± 0.21mm; mean percentage difference $16 \pm 12.5\%$, $p < 0.005$). The LCA-to-aortic annulus (LCA/Ao) and RCA-to-aortic annulus (RCA/Ao) ratio fall within a narrow range that is independent of sex, weight, height or body surface area. The LCA/Ao ratio $= 0.15 \pm 0.02$ (range 0.09 to 0.21), and RCA/Ao ratio $= 0.13 \pm 0.02$ (range 0.09 to 0.20). Conclusion: The regression equations and normograms for the LCA and RCA will serve as useful guides to determine coronary abnormalities, especially in children with Kawasaki Disease. The LCA/RCA-aortic ratio can serve as a quick index to detect coronary dilatation/ectasia.