29

THE RELATION BETWEEN CALCIFICATION AND RESTENOSIS OF CORONARY ARTERY AFTER PERCUTANEOUS TRANSLUMINAL CORONARY ROTATIONAL ABLATION (PTCRA) IN KAWASAKI DISEASE

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Background: Severe calcification in the coronary lesions has been frequently revealed in Kawasaki disease (KD). Therefore, PTCRA has been performed as an effective intervention for obstructive coronary lesions. However, restenosis after PTCRA in KD has not been clear. So, we assessed the relation between calcification and restenosis after PTCRA in KD using the intravascular ultrasound (IVUS) in two KD patients. **Case reports:** Case1; 13 year-old boy, 9 years after the onset of KD, developed 90% stenosis both at AHA segments 2 and 6 on CAG. PTCRA was performed for these stenotic lesions, and consequently stent implantation was added at segment 6 because of intimal dissection after additional PTCA . Stenosis at segments 2 and 6 reduced to 25% and 0%, respectively . IVUS revealed partial circumferential calcification at segment 2, and total calcification at segment 6 . Three months later, the follow-up CAG and IVUS showed 50% restenosis and partial circumferential calcification with intimal thickning at segment 2 . On the other hand, they showed no restenosis and total calcification without intimal thickning at segment 6 . Case2; 16 year-old boy, 14 years after onset of KD, developed 90% stenosis at segment 6, and total occlusion of RCA with recanalization . Stenotic lesion at segment 6 was improved to 50% by PTCRA . Three months later, follow-up CAG and IVUS showed no restenosis and total calcification without intimal thickning . **Conclusion:** PTCRA was performed for three calcified stenotic lesions in 2 KD patients . One, with partial circumferential calcification, caused a restenotic change with developed intimal thickning. On the other hand, other 2 lesions with total calcification did not develop restenosis . These results suggest that total calcification could protect intimal proliferation contributing to coronary restenosis after PTCRA .

30

EFFECTIVENESS AND LUMINAL CHANGES OF ROTABLATOR WITHOUT ADJUNCTIVE PTCA FOR CALCIFIED STENOSIS ASSOCIATED WITH CORONARY ANEURYSM OF KAWASAKI DISEASE

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Background; Highly calcified stenosis associated with coronary aneurysm is often noted in Kawasaki disease.These lesions are difficult to treat by balloon angioplasty or stenting because of their anatomical characteristics. The porpose of this study is to evaluate the efficacy of Rotablator without additional balloon angioplasty to avoid vessel damage. **Method;** 11lesions of 7 patients (LMT 1, LAD 10;12.3 ±2.1yrs;male 6, female 1) were treated by Rotablator without additional balloon angioplasty. All cases were evaluated postoperatevely,3,6,and12months later by angiography and IVUS. **Results;** All procedures were successful. No angiographic or clinical restenosis was noted in the follow-up period (8.6 ±4.4months). Final burr size is 2.0±0.2mm. Angiographic result is summerized as Table1. **Conclusion;** Rotablator without additional balloon angioplasty would be an effective to treat highly calcified stenosis associated with coronary aneurysm in Kawasaki disease,providing no angiographic or clinical restenosis in 12 months after the procedure

Table 1. Angiographic Result

	MLD on IVUS (mm)	% DS
Pre-Op		87.5 ± 11.3
Post-Op	2.1 ± 0.2	42.3 ± 10.4
3-M F/U	2.1 ± 0.2	41.2 ± 9.6
6-M F/U	2.2 ± 0.3	37.9 ± 10.2
12-M F/U	2.1 ± 0.3	39.9 ± 9.4

31

CORONARY ARTERY BYPASS GRAFTING BY UTILIZING ONLY ARTERIAL GRAFTS IN PATIENTS WITH CORONARY ARTERY ANEURYSM AFTER KAWASAKI DISEASE

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Recent studies have reported the efficacy of coronary arterial bypass grafts (CABG) for obstructive lesions of coronary arteries in Kawasaki disease; however, the efficacy in coronary aneurysms is still remains controversial. Recently we studied the coronary flow velocity and perfusion pressure of coronary aneurysms. The results of that study suggested that the stagnation of flow and the reduction of shear stress in large aneurysms could initiate thrombus formation. Thus, we have adopting CABG for patients with large coronary aneurysm to avoid future ischemic damage to the distal myocardium. Furthermore, to achieve long term patency, revascularization was performed using only arterial grafts. Thirteen patients (mean 11.9 years old) with Kawasaki disease underwent CABG in the last 10 years at our hospital. In these patients coronary aneurysms were located at the LMT in 4, LAD in 10, LCX in 4 and RCA in 9. As the arterial grafts, the left internal thoracic artery was used for all patients (8 were to LAD only, 5 were to the diagonal branch and LAD as a sequential graft), whereas the right internal thoracic artery and the right gastroepiploic artery were used for 3 and 1 patients respectively. All the patients survived from the operation without major morbidity. Coronary angiogram showed 100% patency and satisfactory growth of the grafts. Moreover, six of the large aneurysms showed complete degradation during the follow-up period. As a conclusion, we successfully applied CABG using only arterial grafts for the aneurysm as well as the stenosis of coronary arteries without any mortality and morbidity. The long term patency and the growth of the grafts were satisfactory. We consider that CABG with arterial grafts improve the patient's quality of life.

32

REVISION OF DIAGNOSTIC GUIDELINE OF KAWASAKI DISEASE

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Diagnostic guideline of Kawasaki Disease has not been revised since 1984. There has been several progressions during these years concerning management of Kawasaki disease. So a subcommittee for the revision of the diagnostic guideline was organized in the Kawasaki Disease Research Committee to meet the present situation. A draft of new diagnostic guideline was made this year and the final revision will be published next year. Major alterations in this new daft are summarized as follows:1. Cases with 4 or less febrile days shortened by early IVIG treatment were proposed to be equivalent to cases with 5 or more febrile days in the previous criteria. The reason is that many experienced pediatricians have made a diagnosis of Kawasaki disease for the cases even before the 5th febrile day when there were other principal symptoms of Kawasaki Disease. The latest 16th Japanese nationwide surveillance has shown that approximately 10% of patients (total 12,829) started to receive IVGG treatment on or before the 3rd day of illness and 30% of them before 4th day. And also some cases became afebrile before the 5th febrile day when single high dose IVIG was given. 2. The clinical importance of atypical (incomplete) cases is emphasized in the new diagnostic guideline since even atypical cases often developed coronary artery abnormalities and also the incidence of coronary artery abnormality is not significantly different between typical cases and them. 3. The order of 6 principal symptoms of Kawasaki disease was rearranged from head to toe (except for fever and cervical lymphadenopathy) to remember easily: 1. Fever, 2.Conjunctiva congestion, 3.Changes of lips and oral cavity, 4. Rash, 5.Changes of extremities, 6. Cervical lymphadenopathy.

33

KAWASAKI DISEASE: DO WE NEED A NEW CASE DEFINITION?

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Misdiagnosis or missed diagnosis of Kawasaki disease (KD) is a problem in the United States (U.S.). To learn about current practice in diagnosing KD, we conducted a questionnaire survey of general pediatricians in San Diego County and U.S. members of the Pediatric Infectious Disease Society. Responses were obtained from 132/227 (58.1%) generalists and 345/651 (53.0%) subspecialists. Half of the general pediatricians and 90% of the pediatric subspecialists reported having made the diagnosis of KD in febrile patients (pts) who did not fulfill 4/5 clinical criteria. When asked about clinical criteria that trigger consideration of KD, generalists and subspecialists listed fever, rash, and conjunctival injection as the top three clinical signs. Cervical lymphadenopathy was considered an important criterion by the generalists (69/131, 52.7%) but not by the subspecialists (34/334, 10.2%). Despite the well-documented occurrence of KD throughout the pediatric age group, 71/124 (57.3%) generalists and 86/325 (26.5%) subspecialists did not consider the diagnosis in pts <6 mos of age. Similarly, 64/124 (51.6%) generalists and 81/324 (25.0%) did not consider the diagnosis of KD in pts >8 yrs. Laboratory studies reported to be most helpful in establishing the diagnosis of KD were (generalists;subspecialists): erythrocyte sedimentation rate (68.2%;74.2%), complete blood count (58.1%;40.0%), platelet count (51.9%;32.1%), urinalysis (46.5%;41.2%), and echocardiogram (45.0%;52.1%). Education about KD in the U.S. must emphasize that this disease can occur across the entire pediatric age range. Physicians have found the case definition to be overly specific and inadequately sensitive in establishing the diagnosis of KD. Formulation of a new case definition for improved sensitivity in early detection of KD cases may be warranted.

34

SENSITIVITY OF THE KAWASAKI CASE DEFINITION FOR DETECTING CORONARY ARTERY ABNORMALITIES

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Although developed as an epidemiological tool, the Kawasaki Disease (KD) case definition has become a clinical tool used to detect and treat children with KD. Based on our experience treating 103 cases at Children's Hospital, San Diego from January 1998 through December 2000, we found that the Center for Disease Control (CDC) KD case definition results in failure to detect children with coronary artery abnormalities (CAA). Using published nomograms for coronary artery diameters based on body surface area, we determined the prevalence of CAA in all children receiving IVIG treatment for Kawasaki disease at Children's Hospital, San Diego. There were 102 patients with one recurrence. 86/103 cases met the Center for Disease Control case definition (typical KD) and 17/103 did not (atypical KD). CAA occurred in 42/86(49%) typical KD cases and 10/17 atypical KD cases (59%). These data demonstrate that 50%(52/103) of KD patients have detectable CAA. Five patients developed coronary aneurysms- 2 in the atypical group (12%) and 3 in the typical group (3.5%). The sensitivity of the KD case definition for detecting children with CAA was only 0.81(42/42+10). With this level of sensitivity for CAA detection, reliance on the CDC case definition as the sole clinical tool for diagnosis results in physician failure to detect children with CAA. Because there is an effective treatment, clinicians need a case definition that is highly sensitive. As the treatment is relatively safe, specificity of the case definition is less important. Efforts to revise the clinical case definition in order to improve its sensitivity for CAA are warranted. New criteria may include clinical laboratory findings such as leukocyte count or ESR. Revisions may eliminate current criteria such as cervical lymphadenopathy. Candidate clinical case definitions should undergo prospective testing with CAA detection as the primary outcome.