

Coronary Artery Bypass Graft in Kawasaki Disease Patients: Siriraj Experience

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Kawasaki disease (KD) is an acute systemic vasculitis. It is one of the most common acquired heart diseases in children. Its important acute complication is coronary artery aneurysm. Although most of these aneurysms resolved overtime but some never did. Coronary artery stenosis and myocardial insufficiency or infarction are late complications. Coronary artery bypass graft (CABG) and catheter intervention are the treatment for these patients. We report our first five cases of Kawasaki patients with myocardial insufficiency who underwent CABG at Siriraj Hospital. Patients' ages ranged from 3.3 to 14.4 years old. Only two patients (40%) had angina. Internal thoracic artery was used as a bypass graft in most patients and postoperative course was uneventful. Coronary bypass operation is a safe and reliable surgical modality for coronary artery sequelae in children with Kawasaki disease. A long-term follow-up is necessary to study the long-term outcome of bypass.

Keywords: Kawasaki disease, Coronary artery bypass graft (CABG)

J Med Assoc Thai 2005; 88(Suppl 8): S197-202

Full text. e-Journal: <http://www.medassocthai.org/journal>

Kawasaki disease (KD) is an acute systemic vasculitis, first described by Dr. Kawasaki in Japan in 1967⁽¹⁾. It is one of the most common acquired heart diseases in children. Most of the children with Kawasaki disease are less than five years old with majority occurring in oriental children. The etiology of this disease is unknown. Recently some viral infection and genetic polymorphisms have been implicated⁽²⁾. Diagnosis of this disease is made by clinical criteria and some clinical and laboratory findings^(3,4). Its important acute complication is coronary artery aneurysm. The treatment of KD includes infusion of high dose immunoglobulin in order to reduce risk of coronary aneurysms. Such aneurysms occurred in 15% to 25% of untreated patients⁽⁵⁾ but some of treated patients still have this complication (3% to 5%)^(6,7). Although

most of these aneurysms resolved overtime but some never did, especially for those with giant aneurysms (diameter of aneurysm over than 8 mm). Patients with these complications might developed coronary artery stenosis, myocardial infarction and sudden death⁽⁸⁾. Other interventions such as cardiac catheterization with intervention or coronary revascularization surgery have treatment role in these patients.

During the last 5 years, coronary artery bypass graft (CABG) was employed in 5 Kawasaki patients with coronary abnormalities at Siriraj Hospital with good result. Post-operatively, all of them had New York Heart Association (NYHA) I and had good quality of life.

Material and Method

We conducted a retrospective chart review of children with Kawasaki disease who underwent coronary artery bypass grafts and were diagnosed at Siriraj Hospital from January 2000 to December 2004. Clinical

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records were reviewed for the followings: patient demographics, initial presentation of KD, initial laboratory findings, initial treatment, initial and follow-up echocardiographic findings, others investigations such as perfusion scan and/or exercise stress test, coronary angiography, symptoms of chest discomfort, duration from initial diagnosis to surgical procedure, surgical procedure and postoperative results.

Results

We identified 5 patients who were diagnosed KD at Siriraj Hospital from 1990 to 2004 who underwent coronary bypass graft (CABG) within last five years. These five patients forms the basis of these reports.

Clinical data

During the last 5 year, we performed coronary artery bypass graft in 5 patients with Kawasaki disease and coronary complications at Siriraj Hospital. Patients' ages range from 3.3 to 14.4 years with mean age of 7.2 ± 4.4 years. The age at initial diagnosis of Kawasaki disease ranged from 1 ± 2.9 years with mean age of 1.98 ± 0.8 years. Three patients (60%) were referred from other hospitals. Two of them (40%) was diagnosed as cases with incomplete Kawasaki disease (3/5 criteria) and all of them had elevated erythrocyte sedimentation rate (over 90 mm/hr). Single dose of 2 gm/kg of IVIG was used in 3 patients. One patient received 2 doses of 2 gm/kg of IVIG and the other did not receive IVIG because he was referred too late (18th day of fever). Three of the patients received IVIG after the 10th day of fever because of the delayed diagnosis of the disease.

Clinical data (Table 1)

Echocardiogram was done on the day of initial diagnosis and most of them (60%) had abnormal coronary arteries. All had abnormal coronary findings by echocardiogram after two months of the disease. Four (80%) had other investigations such as myocar-

dial perfusion scan or cardiac MRI in order to evaluate myocardium perfusion. Coronary angiogram was performed in all patients and all had both right and left coronary abnormalities.

Cardiac data (Table 2)

The duration from the diagnosis of KD to CABG ranged from 11 to 160 months (mean = 62.40 ± 58.85 months). Only two patients had symptoms of chest discomfort and these symptoms did not correlate with results of myocardial perfusion scan.

Numbers of bypass graft ranged from 1 to 3 lesions (mean = 2 ± 0.7). Left internal thoracic arteries were used as grafts for 4 lesions, right internal thoracic arteries for 2, radial artery for 1 and saphenous vein for 1 lesion.

Postoperative evaluation

No perioperative and post operative death was observed. All had NYHA 1 and did not have any symptoms of myocardial ischemia. Echocardiogram showed normal left ventricular ejection fraction in 4 patients (80%). Post operative coronary angiogram was done in one patient at 1 year after surgery. The result showed graft patency.

Discussion

Kawasaki disease is one of the most common acquired heart diseases in childhood. It is an acute vasculitic illness of young children ages less than five years. It typically presents with prolonged fever, bilateral non exudative bulbar conjunctivitis, mucous membrane changes, skin rash, swelling or erythema of hands and feet and unilateral cervical lymphadenitis⁽⁴⁾. There is no reliable laboratory test for the diagnose this disease, and thus diagnosis is often made by recognition of clinical signs consistent with Kawasaki diagnosis criteria and exclusion other diseases.

Kawasaki is self-limited disease but its complication can be serious. Long-term treatment of this

Table 1. Clinical data of the five patients who underwent CABG

| Patients | Gender | Age | Day of fever at initial treatment | Criteria | ESR |
|----------|--------|-----|-----------------------------------|----------|-----|
| A | male | 2.9 | 3 | 4/5 | 90 |
| B | male | 1.0 | 17 | 3/5 | 144 |
| C | male | 1.2 | 12 | 3/5 | 110 |
| D | male | 2.0 | 18 | 5/5 | 109 |
| E | male | 2.8 | 5 | 5/5 | 92 |

Table 2. Cardiac data of the five patients in this study

| Patients | Echocardiogram | Perfusion scan or Cardiac MRI | Angiogram | Surgery | Symptoms of chest discomfort |
|----------|--------------------------------------|-------------------------------|---|---|------------------------------|
| A | Normal (day6); LAD and RCA aneurysm | No | RCA aneurysm with long segment ectasia, LMCA aneurysm | double graft, LIMA to LAD, Free graft of RIMA to PD | Yes |
| B | LAD and RCA aneurysm | Both | LAD aneurysm, mild stenosis post aneurysm. RCA aneurysm | LIMA to LCA, RIMA to RCA | No |
| C | LMCA, LCx and RCA aneurysm | Cardiac MRI | LCx aneurysm, ectasia of LMCA and LAD and RCA aneurysm | LIMA to LAD, RIMA to RCA and lt radial to LCx | No |
| D | Normal (day18); LAD and RCA aneurysm | Perfusion scan | RCA aneurysm with proximal stenosis, multiple stenosis of LAD | LIMA to LAD, aneurysmorrhphy of aneurysm of RCA | Yes |
| E | LMCA, LAD and RCA aneurysm | Cardiac MRI | RCA aneurysm, LAD and LCx aneurysm | LIMA to LAD and SVG to PD | No |

LMCA = left main coronary artery, LAD = left anterior descending coronary artery, LCx = left circumflex coronary artery, RCA = right coronary artery, LIMA = left internal thoracic artery, RIMA = right internal thoracic artery, SVG = saphenous vein graft

disease depends on remaining coronary artery status⁽⁹⁾. Although incidence of serious coronary complication is fortunately low but if occurs, such as myocardial infarction, the mortality rate is very high. In our study, symptoms of the patients, especially in small children, did not correlate with the degree of myocardial ischemia. Thus, it is possible to misdiagnose these important problems in these patients.

The diagnosis of Kawasaki disease is delayed in some of our patients. This problem could be explained by multiple factors such as incomplete presentation of criteria of the disease, misdiagnosis by the doctor, poor socio-economic and educational status of the patients⁽¹⁰⁾. Such problems affect long-term outcome of patients because delayed diagnosis patients have more coronary complications than early diagnosis group. No study confirmed that treatment of IVIG after 10th day of illness has any benefit but the guideline from

American Heart Association suggests that IVIG also should be administered to children presenting after the 10th day of illness if they have either persistent fever without other explanation or aneurysms and ongoing systemic inflammation, as manifested by elevated ESR or CRP⁽⁴⁾. In our study, 3 patients (60%) who underwent CABG were diagnosed after 10th day of illness and although 2 of them received IVIG, coronary complication could not be prevented.

Patients with KD who had coronary complications should be followed regularly because coronary artery lesions could change dynamically with time. Serial stress tests and myocardial imaging are mandatory in the management of these patients so as the need for coronary angiography and for surgical or transcatheter intervention can be determined. This study reveals that the duration from initial diagnosis to the time of surgery varies, i.e., range from 11 to 160

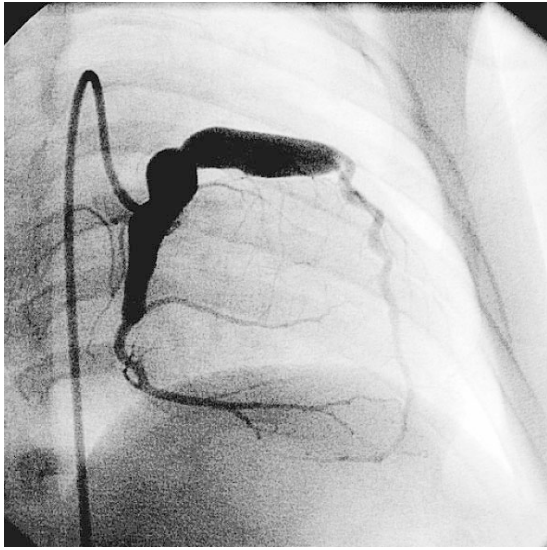


Fig. 1.1 Left coronary angiogram showing giant aneurysms of left main coronary artery (LMCA), left anterior descending (LAD) and left circumflex coronary artery (LCx).

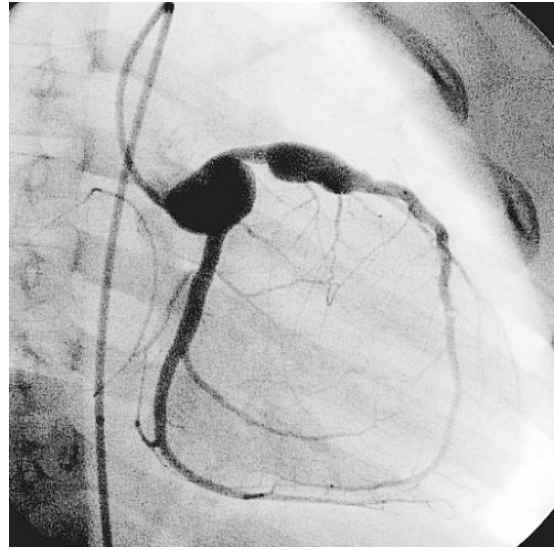


Fig. 1.2 Left coronary angiogram of the same patient, one year later, showing regression of aneurysms. The patient still had LMCA and LAD aneurysms.

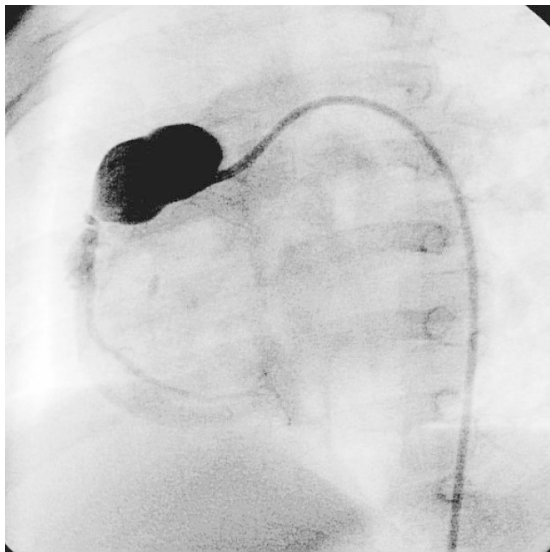


Fig. 2.1 Right coronary angiogram showing giant aneurysm of proximal right coronary artery (RCA).

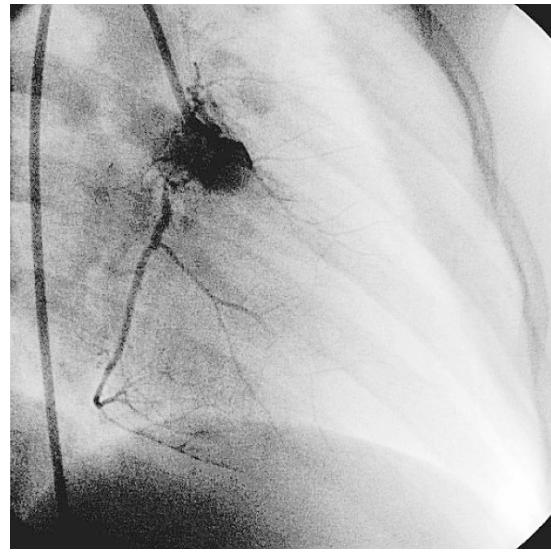


Fig. 2.2 One year later, the aneurysm regressed and stenosis of proximal RCA was observed.

months. Patients with giant aneurysm should be educated about their risk although they did not have any symptoms for years.

The indication for catheter interventions or surgical procedure (CABG) is controversial. The indication for CABG depends not only by findings derived

from coronary angiography, but also on other clinical factors, such as severity of myocardial ischemia, history of myocardial infarction and ventricular performance. In some study, aneurysm alone was an indication of surgery if there was any kind of ischemic event⁽¹⁾.

Saphenous vein graft (SVG) was first used in CABG for this disease in 1976 by Kitamura⁽¹²⁾. They later reported the efficacy of internal thoracic artery (ITA) as a bypass graft in pediatric patients in 1985⁽¹³⁾. ITA is more acceptable than SVG as a bypass graft in children since patency rate of ITA in longer term is better than SVG. The patency rates of the arterial grafts were 94, 82 and 78% at 1, 5 and 10 years, respectively, and this was higher than that of the venous grafts (82, 63 and 36%, respectively)⁽¹⁴⁾. Presumably, degeneration of SVG progresses more frequently and more rapidly. The arterial graft (ITA) can grow both length and diameter but SVG may not. Use of SVG should be avoided unless ITA is unavailable. Other arterial grafts that can be use in childhood other than ITA are right gastroepiploic and radial artery⁽¹⁵⁾.

For CABG in KD at Siriraj Hospital, we preferred ITA and if more grafts are needed then we would use radial artery or SVG. The long-term patency of these graft should be closely followed.

Other choices of treatment of KD patients with coronary artery stenosis are transcatheter intervention. The indication for catheter interventions is not similar to CABG. Coronary artery bypass graft (CABG) is best indicated in those with critical stenosis or entire occlusion of the coronary arteries, while catheter intervention is indicated in those with less critical obstructions. Catheter interventions can be repeatedly employed. Sugimura reported that PTCRA was of use for recanalizing the coronary arteries with severe calcification⁽¹⁶⁾. We do not have experience for this kind of intervention for KD patients at our hospital.

Most studies indicated that KD patients had good outcome after surgery⁽¹⁷⁾. They had improvement of ejection fraction of left ventricle and had good quality of life when compared to pre surgery condition. The results in our patients confirmed such previous findings.

Conclusion

KD is one of the most common acquired heart diseases in childhood. Primary physician should be aware of this disease in young children with history of prolonged fever more than 5 days. Early treatment before the 10th day of the disease is important because the appropriate treatment can minimize coronary complication. KD patients with coronary aneurysm should be followed regularly for early detection of myocardial insufficiency from coronary stenosis. CABG can use for treating these patients with good result. Long-term follow-up is required in these patients.

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การผ่าตัดแก้ไขหลอดเลือดหัวใจตีบในผู้ป่วยโรคคาวาซากิในโรงพยาบาลศิริราช

ประคัลภ์ จันทร์ทอง, สมชาย ศรียศชาติ, กฤตย์วิกรม ดุรงค์พิศิษฐ์กุล, จารุพิมพ์ สูงสว่าง, ดวงมณี เลหาประสิทธิ์พร, อภิชาติ นานา

โรคคาวาซากิเป็นโรคที่มีการอักเสบของหลอดเลือดทั่วร่างกายโดยเฉียบพลัน พบได้บ่อยในเด็กเล็ก และมีภาวะแทรกซ้อนที่สำคัญคือ เส้นเลือดที่เลี้ยงหัวใจ (coronary artery) โป่งพอง โดยส่วนหนึ่งของความผิดปกตินี้จะหายไปได้เอง แต่ในผู้ป่วยบางรายอาจเกิดภาวะเส้นเลือดเลี้ยงหัวใจตีบและเกิดภาวะกล้ามเนื้อหัวใจขาดเลือดตามมาได้ การรักษาภาวะดังกล่าวทำได้โดยการผ่าตัดแก้ไข (coronary artery bypass graft) หรือการรักษาโดยการสวนหัวใจ การศึกษานี้รายงานการรักษาผู้ป่วยโรคคาวาซากิที่มีภาวะเส้นเลือดเลี้ยงหัวใจตีบที่โรงพยาบาลศิริราชโดยการผ่าตัดรักษาจำนวน 5 ราย โดยมีอายุของผู้ป่วยตั้งแต่ 3.3 ถึง 14.4 ปี โดยร้อยละ 40 ของผู้ป่วยมีอาการเจ็บหน้าอกจากกล้ามเนื้อหัวใจขาดเลือด (angina) การผ่าตัดส่วนใหญ่ (ร้อยละ 75) ใช้เส้นเลือด internal thoracic artery ในการแก้ไข ผลการรักษาผู้ป่วยทั้งหมดได้ผลดี และควรมีการติดตามผู้ป่วยเหล่านี้ต่อไปในระยะยาว