Aortocoronary Bypass Grafting in a Child with Coronary Artery Obstruction Due to Mucocutaneous Lymphnode Syndrome

Report of a Case

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SUMMARY A four-year-old boy with a myocardial infarct and total occlusion of the right coronary and the left anterior descending coronary arteries due to mucocutaneous lymphnode syndrome (MCLS), confirmed by selective coronary arteriography, underwent successful double aortocoronary bypass grafting. Patency of the grafts was demonstrated by graft angiography and the improvement of the contractile pattern of the left ventricle was reflected by the increase in ejection fraction from 0.45 to 0.61. This is the first patient successfully treated by aortocoronary bypass grafting for coronary artery obstructive lesions due to MCLS.

This experience has demonstrated the feasibility of surgical management in a child with coronary artery obstruction due to MCLS. However, there are unanswered questions regarding the fate of the saphenous vein graft in relation to the growth of a child. Long-term clinical and angiographic follow-ups are mandatory to determine the significance of this mode of surgical treatment for the sequel of MCLS.

MUCOCUTANEOUS LYMPHNODE SYNDROME (MCLS) is a febrile disease prevailing among children in Japan which was first reported as a clinical entity by Kawasaki in 1967. Since then, more than 7,000 patients have been reported and the number of patients is still increasing. MCLS has come to the attention of pediatric cardiologists since cardiac or sudden death can occur either during or long after the acute phase of the illness. Autopsy studies have always demonstrated the presence of coronary artery lesions such as aneurysm formation and/or thrombotic obstruction. The cause of death in some patients was apparently myocardial infarction. Even among the surviving patients, it has now become evident from angiographic studies that coronary artery lesions are present with an incidence as high as 60 to 80. The real etiology of MCLS remains uncertain and no definitive treatment has been established for coronary artery obstruction as a sequel of MCLS in children. This paper describes the first attempt at surgical treatment for coronary artery obstructive lesions due to MCLS in a child.

Case Report

A 4-year-old Japanese boy had been well until May 23, 1974, when he developed a high fever up to 40°C that did not respond to antibiotics or the usual antipyretics. On the third febrile day, macular skin rash, deviated of the eyeballs, and swelling of the cervical lymphnodes were noted, all of which appeared to be the typical signs of MCLS. The fever lasted for a week and all the signs and symptoms subsided spontaneously. He was discharged on the 35th hospital day in good condition and with no report of electrocardiographic abnormalities.

On March 17, 1975, approximately 10 months after the acute onset of MCLS, he suddenly groaned while taking a nap. Cyanosis, upward deviation of the eyeballs, stretching of the extremities and loss of consciousness occurred. He was hospitalized in a nearby neurosurgical clinic, where he was suspected to have cardiac arrest or ventricular fibrillation with spontaneous recovery. Consciousness returned on the next day and there were no residual neurologic deficits. Definitive electrocardiographic (ECG) abnormalities suggestive of acute myocardial infarction were noted. After ten days he was discharged and referred to the Osaka University Hospital for a more definitive cardiovascular evaluation.

He was a well-developed, well-nourished boy who looked normal. After recovery from myocardial infarction, he had no severe subjective symptoms except for occasional upper abdominal pain which was relieved by nitroglycerin. The chest X-ray revealed a slight enlargement of the heart with a cardiothoracic ratio of 57%. Laboratory studies showed normal values for blood, blood chemistry, liver and renal functions. Blood sedimentation rate was normal and C-reactive protein test was negative, confirming the absence of continual inflammatory process at this time. The ECG exhibited the presence of old anteroseptal infarction (fig. 1a).

On June 26, 1975, cardiac catheterization, left ventricular cineangiography and selective coronary arteriography were performed in this order through the femoral artery and vein. Special catheters for selective coronary arteriography for a child were handmade using a #5 Kifa red catheter after the shape of Judkins catheters. The heart rate

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CORONARY ARTERY ANEURYSM & OBSTRUCTION DUE TO MCLS

April 8, 1975. 4 months before surgery

September 5, 1975. 1 month after surgery

FIGURE 1. Pre (a) and postoperative (b) electrocardiograms. Abnormalities suggestive of old anteroseptal infarction were seen before surgery. Following bypass surgery, there were no significant electrocardiographic changes, but slight improvements of ST-T segments in lead I and V3 through V6 were discernible.

at the time of the study was 96/min in normal sinus rhythm. As summarized in table 1, the left ventricular end-diastolic pressure (LVEDP) was not elevated. The pulmonary artery (PA) pressure was also normal.

Preoperative Coronary Arteriographic Findings

As shown in figures 2a and 2c, the right coronary artery (RCA) was totally obstructed 1 cm from its origin. The distal RCA was again patent in its diaphragmatic portion, bridged by intra and intercoronary collateral networks. The left anterior descending coronary artery (LAD) was also totally occluded at its origin and there was no antegrade blood flow to the LAD (fig. 2 b, c). A small aneurysm was present in the circumflex artery (CXA), but no obstructions existed. The CXA was the only patent main artery allowing blood to perfuse the distal RCA and LAD retrogradely through well-developed collateral channels. The diameters of the coronary arteries distal to the obstruction, visualized through the collaterals, measured approximately 1.5 and 1.0 mm in the RCA and LAD, respectively.

Preoperative Left Ventriculographic Findings (fig. 3)

The cine left ventriculography was performed with the patient in the right anterior oblique position. Hypokinesis was obvious at the anteropapical and inferopapical areas. The end-diastolic volume (EDV), measured by using an ellipsoid formula, was 64 ml (97 ml/m²), a slight enlargement. The end-systolic volume (ESV) was 35 ml (53 ml/m²). The ejection fraction (EF) was computed to be 0.45, a mild depression.

Although he had only occasional anginal pain, his previous history of myocardial infarction complicated by transient circulatory arrest, and the fact that two of the major coronary arteries were totally occluded at their origin, led us to recommend surgery. Angiography demonstrated that the internal mammary artery was too small for a
successful anastomosis. For these reasons, we decided to use the autologous saphenous vein as a graft.

Operation

Operation was performed on August 5, 1975, approximately five months after myocardial infarction. The heart was slightly enlarged and a hypokinetic area was observed at the anteroapical area of the left ventricle. With hypothermic (29°C) cardiopulmonary bypass, the first graft was placed at the diaphragmatic portion of the RCA and the second at the mid-portion of the LAD. The diameters of the coronary arteries at the anastomotic sites measured by probes were approximately 1.5–2.0 and 1.0–1.5 mm in the RCA and LAD, respectively. The walls of the arteries were thin and pliable and looked nearly normal. The CXA was left untouched. After completion of all anastomoses, the patient came off cardiopulmonary bypass easily. The mean graft flow measured just prior to closure of the chest by the use of a Statham electromagnetic flowmeter was 35 ml/min in the RCA graft and 30 ml/min in the LAD graft (fig. 4). Postoperative convalescence was smooth and satisfactory. Anticoagulation therapy with coumadin was started from the third postoperative day. A month after surgery the patient gained full recovery and could resume nearly normal activities. The ECG at this period exhibited no remarkable changes compared to the preoperative one. Postoperative catheterization was carried out one month after surgery. There were no significant changes in heart rate, arterial blood pressure, LVEDP and PA pressure following bypass grafting (table 1).

Postoperative Coronary and Graft Angiographic Findings

As illustrated in figure 5a and c, the graft to the RCA was widely patent, delivering blood to the posterior descending coronary artery. The RCA was found to be a dominant vessel. Distal to the anastomosis, there was no obstruction

Figure 2. (Top left) Preoperative right coronary arteriogram in the left anterior oblique (LAO) position exhibiting total obstruction of the artery near the origin. The artery was again patent from the diaphragmatic portion, connected by intracoronary collateral bridges. (Bottom left) Preoperative left coronary arteriogram showing total occlusion of the left anterior descending artery at the origin, being perfused in a retrograde fashion from the circumflex artery through apical collateral connection. The peripheral right coronary was also opacified through the dilated collateral pathway from the posterior branch of the circumflex artery (intercoronary collaterals). The circumflex artery was the only patent main artery and had a small aneurysm in the main trunk. (Right) Drawings of the coronary arteriographic findings. 1. Total obstruction of right coronary artery (RCA). 2. Reopening of RCA. 3. Right ventricular branch. 4. Posterior descending artery. 5. Small aneurysm of circumflex artery. 6. Circumflex artery. 7. Collateral vessel to left anterior descending artery (LAD). 8. LAD (retrograde filling). 9. Total obstruction of LAD. 10. Diagonal branch. 11. Collateral vessel to RCA. 12. RCA.
or aneurysm formation in the RCA system. The RCA proximal to the anastomosis was also retrogradely perfused up to the portion obstructed. The graft to the LAD was also widely patent allowing blood to perfuse large anterior and septal areas of the left ventricle in both ante- and retrograde fashions (fig. 5 b, c). Many septal perforators and the diagonal branch were now retrogradely perfused.

Angiography of the original coronary arteries was also carried out. The RCA became very small, only perfusing the right atrial branches. By left coronary artery injection, only the CXA was visualized. The small aneurysm of the CXA remained the same and the rich collateral vessels to the distal RCA and LAD opacified prior to surgery were no longer seen following bypass grafting. Blood flow to the RCA and LAD depended entirely on the two grafts inserted.

Table 1. Catheterization and Angiocardio graphics Data

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<th>Abbreviations: HR = heart rate; AO = aortic pressure syst./diast. (mean); LVEDP = left ventricular end-diastolic pressure; PA = pulmonary artery pressure; EDV = end-diastolic volume; ESV = end-systolic volume; SV = stroke volume; EF = ejection fraction; ΔV/ΔP/ESV = mean diastolic compliance (ΔV: volume change in diastole; ΔP: pressure change in diastole).</th>
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<td>Before surgery</td>
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<td>(June 26, 1975)</td>
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<td>HR (beats/min)</td>
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<td>ΔV/ΔP/ESV (mm Hg⁻¹·10⁻⁴)</td>
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Figure 3. Pre (left) and postoperative (right) left ventriculograms taken at the 30° right anterior oblique position. Left) Hypokinesis was observed at the anteroapical-inferoapical wall of the left ventricle before operation. Right) Postoperatively, the contractile pattern of the left ventricular wall markedly improved with an increase in ejection fraction from 0.45 to 0.61.

Figure 4. Simultaneous recordings of aortic pressure and graft flow. Mean graft flows of 35 and 30 ml/min were recorded in the SVG to the RCA and LAD, respectively. The aortic pressure was 82/48 mm Hg, equivalent to the pre and postoperative aortic pressure at catheterization.
Postoperative Left Ventriculographic Findings (fig. 3)

Hypokinesis at the anteroapical and inferoapical walls markedly improved after successful bypass grafting. The pre and postoperative left ventricular functions were compared in table 1. The postoperative EDV was calculated to be 67 ml (101 ml/m²), not significantly different from the preoperative value. The ESV, however, decreased from 35 to 26 ml and the left ventricular stroke volume increased from 29 to 41 ml. The heart rate was equivalent before and after surgery, 96 and 90/min, respectively. The EF, therefore, increased to 0.61 from 0.45, almost normalized after operation. The left ventricular mean diastolic compliance, expressed as \( \Delta V/\Delta P/\text{ESV}^{10} \) also increased following revascularization of the myocardium (\( \Delta V \): volume change in diastole, \( \Delta P \): pressure change in diastole). The patient is now in excellent condition five months after surgery.

Discussion

Although the mortality of MCLS is not high (1 to 2%), the total number of patients in Japan is now estimated to be more than 7,000 and, therefore, more than 100 patients have died from the disease. Although MCLS is prevalent among Japanese children, it is now receiving attention in the U.S.\(^9\). Coronary artery lesions reported as a sequel of infantile polyarteritis nodosa in the U.S.\(^11, 12\) are also quite similar to those of MCLS in Japan. Death from MCLS can occur not only in the acute phase of the illness, but also long after it subsides.\(^6\) The cause of death was always attributed to aneurysm and thrombotic obstruction of the coronary artery. However, there has been no definitive treatment for this catastrophic sequela of this disease. We have previously reported a successful surgical experience with a 26-year-old man having multiple coronary artery aneurysms and obstruction due most probably to arteritis of a similar type.\(^13\) Based upon this experience, we decided to treat this 4-year-old patient by the same method. He is the first treated by aortocoronary bypass grafting for coronary artery obstructive lesions due to MCLS.

Although aortocoronary saphenous vein bypass grafting has become an established surgical mode of therapy for arteriosclerotic coronary artery disease in adults, questions arise as to the fate of the SVG inserted in childhood. The mean blood flow measured during operation was only 30 to 35 ml/min in each graft, but this seemed to be adequate for the 4-year-old child's heart. The volume of the left ven-

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**Figure 5.** Saphenous vein graft (SVG) angiograms. (Top left) The SVG to the RCA was patent allowing blood to fill the posterior descending artery. The RCA was a dominant artery. There was no abnormality observed in the distal RCA. Retrograde filling of the RCA to the obstructed portion was also seen. (Bottom left) The SVG to the LAD was also patent, allowing blood to flow in both anterograde and retrograde fashions. Many septal perforators and the diagonal artery were visualized. (Right) Drawings of the SVG angiographic findings. 1. SVG to RCA. 2. Anastomotic site. 3. Retrograde filling of RCA. 4. Posterior descending artery. 5. SVG to LAD. 6. Anastomotic site. 7. Distal LAD. 8. Proximal LAD (retrograde filling). 9. Diagonal branches. 10. Septal branches.
tricular muscle to be perfused appeared to be less than one third that of an adult, judging from the heart size. The angiographic diameters of saphenous vein grafts were measured to be 3.0 and 2.5 mm in the RCA and LAD grafts, respectively. The diameters of these grafts were larger than that of the IMA in adults. Accordingly, there seems to be no problem with regard to the diameter of the graft even when the patient grows into adulthood. However, the question concerning the length of the SVG related to the patient’s growth remains unanswered. Chiariello et al. reported 6- and 11-year-old children with anomalous origin of the left coronary artery from the pulmonary artery treated by SVG insertion. These patients had well patent grafts 3.5 and 7 years after operation. Although the grafts appeared to be much longer in our patient than those in patients treated for anomalous origin of the left coronary artery, stretching of the graft may not occur if the uninvolved distal portions of the coronary arteries grow in such a manner as to compensate for the limited length of the grafts. Whether or not the patient may need reoperation in the future cannot be determined at present.

From the pathologic viewpoint, MCLS is an arthritis of a special type involving small and medium sized muscular arteries. Onouchi et al. reported that MCLS has a predilection for involvement of the main coronary arteries due to nutrient arteritis of the large coronary artery. From their observation and our surgical experience, bypass grafting may be the treatment of choice for severe coronary artery lesions due to MCLS in selected patients. Further detailed evaluation of coronary artery lesions in MCLS is certainly essential.

In our patient, the contractile pattern of the left ventricle improved and the ejection fraction became almost normalized after successful myocardial revascularization. He has now three coronary artery systems compared to one prior to surgery. We believe that the surgical treatment improved his left ventricular function and reduced the possibility of further myocardial infarction. However, because of the undetermined fate of aSVG inserted in childhood, long-term clinical and angiographic follow-ups are mandatory and a surgical indication should be cautiously made at the present time.

References

Aortocoronary bypass grafting in a child with coronary artery obstruction due to mucocutaneous lymphnode syndrome: report of a case.
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