# Long-Term Outcomes of Pediatric Coronary Artery Bypass Grafting and Down-Sizing Operation for Giant Coronary Aneurysms

**Yuji Maruyama and Masami Ochi**

**Abstract** There are several concerns regarding surgical revascularization for Kawasaki coronary disease, including the choice of conduit, optimal timing, and indications for coronary artery bypass grafting (CABG). The internal thoracic artery is the best conduit for pediatric CABG because of its favorable growth potential and long-term patency. Use of a saphenous vein graft should be avoided unless the internal thoracic artery is unavailable. Indications for CABG for Kawasaki coronary disease have not yet been established. In principle, coronary aneurysms should be observed continuously for 1–2 years under restrictive anticoagulation therapy, because coronary aneurysms regress in 50 % of patients within 1–2 years. Presence of severe ischemia with giant coronary aneurysms involving obstructive lesions of the left main trunk or left anterior descending artery (LAD) is an unequivocal indication for CABG. In addition, a giant aneurysm with recurrent thrombosis under restrictive anticoagulation therapy or with severely delayed flow without significant localized stenosis may be an indication for CABG. However, determining surgical indications is difficult, especially for younger children, because of technical challenges. To prevent fatal complications, CABG might be indicated at a young age for patients with severe ischemia, because a history of myocardial infarction and impaired cardiac function affect prognosis. Down-sizing operation for giant aneurysms of non-LAD lesions without significant stenosis and severe calcification may be a good choice to improve coronary circulation and allow discontinuation of warfarin, if indications for this procedure can be established.

**Keywords** Coronary artery bypass grafting • Down-sizing operation • Internal thoracic artery • Giant coronary aneurysm

Y. Maruyama (✉) • M. Ochi
Department of Cardiovascular Surgery, Nippon Medical School, 1-1-5 Sendagi, Bunkyo-ku, Tokyo 113-0022, Japan
e-mail: maruyamayuji@nms.ac.jp



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## Long-Term Outcomes of Pediatric CABG

### Introduction

The long-term outcomes for coronary artery bypass grafting (CABG) for patients with Kawasaki disease (KD) remain uncertain. Issues involved in improving long-term outcomes for pediatric CABG include conduit choice and the optimal timing and correct indications for CABG. These issues will be discussed in the next section.

The characteristics of pediatric CABG need to be considered for patients with Kawasaki coronary disease. KD coronary artery lesions are quite different from those seen in adult atherosclerotic disease. First, patients with atherosclerotic coronary disease occasionally have multivessel and/or diffuse stenotic lesions and require multivessel revascularization, whereas patients with Kawasaki coronary disease usually have coronary aneurysms and subsequent obstructive changes in only the proximal portion of the left anterior descending artery (LAD) or right coronary artery (RCA) and require 1- or 2-vessel revascularization. Second, the mean age of patients with atherosclerotic disease undergoing CABG is 60–70 years, whereas that of patients undergoing pediatric CABG is around 10 years. Hence, patients undergoing pediatric CABG have a longer life expectancy, and quality-of-life considerations are more important than for adult patients. Third, off-pump technique is not applicable in pediatric CABG because young patients do not have the usual risk factors for cardiopulmonary bypass, such as diabetes or failure of other organs, including cerebrovascular disease, peripheral vascular disease, or chronic kidney disease. Reliable conduits with expected long-term patency thus need to be anastomosed accurately under on-pump arrest conditions in pediatric CABG.

### Choice of Conduit

Kitamura et al. reported the first pediatric CABG using the saphenous vein graft (SVG), in 1976, [1] and the internal thoracic artery (ITA), in 1985 [2]. Their results showed a significantly higher actuarial patency rate for the ITA than for the SVG, and this difference was even greater in young children [3–7]. They reported that the actuarial patency of SVG for patients aged $\leq$10 years was extremely poor, 25 % at 3 years postoperatively, and most patients with patent SVG had various degrees of degenerative change [3]. Moreover, late cardiac death was strongly related to absence of an ITA graft [4, 5]. They therefore recommended avoiding the use of SVG unless the ITA is unavailable [3–6].

The ITA appears to be the best conduit for pediatric CABG because of its growth potential, excellent long-term patency [8], and biological characteristics [9]. A Japanese national survey of multiple centers found that ITA patency for patients aged ≤12 years was less favorable than for those >12 years, and this difference appeared to be mainly due to technical difficulties encountered in younger children [7]. Moreover, late death was strongly related to younger age at the time of surgery, and the absence of ITA grafts, in a previous multicenter study [4]. However, Kitamura et al. later showed that ITA patency did not depend on patient age at surgery [3]. We believe that left internal thoracic artery (LITA)-to-LAD grafting is the gold standard even in children >1 year of age or >10 kg in body weight.

Use of the right internal thoracic artery (RITA) is recommended as a second arterial conduit. Kitamura et al. first reported bilateral ITA use for pediatric CABG in 1990 [10] and concluded that it should be used whenever indicated [3, 4, 10]. The superiority of bilateral ITA over single ITA was confirmed in adult patients over a 10-year period after CABG [11]. KD patients may benefit much more than adult patients from bilateral ITA grafting because of their longer life expectancy. However, the usefulness of RITA is less than ideal because of the short length of the ITA graft available from the flat, short chest of young children. Moreover, the RITA is often not long enough to reach the LAD, left circumflex artery (LCx), or distal RCA, even in adult patients. A Y-composite graft using bilateral ITA grafts is useful for multivessel CABG in adult patients [12]. However, mild stenosis of the native coronary artery is a significant predictor of competitive flow and graft occlusion for Y-composite grafts [13]. In KD patients, evaluating the degree of localized stenosis around giant aneurysms may be difficult because of severe flow turbulence [14]. A complicated graft design, such as the Y-composite graft, should therefore be avoided in pediatric CABG for Kawasaki coronary disease.

The right gastroepiploic artery (GEA) was first used for pediatric CABG in 1990 [15] and has also been used recently by others, with favorable early results [3]. However, the long-term patency of GEA in young children has not been assessed in a large series. Routine use of the GEA may not be practical for young children because its size depends on the patient. In addition, the risk of flow competition is greater for the GEA than for the ITA because the GEA is the fourth branch of the abdominal aorta. The GEA can thus only be used in large, older children. The radial artery (RA) has been used only in redo cases [3], and long-term patency of the RA has not been assessed in pediatric CABG.

## *CABG Timing and Indications*

The indications for CABG in Kawasaki coronary disease were described in the "Guidelines for diagnosis and management of cardiovascular sequelae in Kawasaki disease (JCS 2008) [16]" (Table 1), which is based on a guideline published two decades ago [17]. CABG is considered when there are (1) severe occlusive lesions in the left main trunk (LMT), (2) severe occlusive lesions in multiple vessels, (3) severe occlusive lesions in the proximal portion of the LAD, or (4) jeopardized collaterals. In addition, indications for younger children are described [16]. Whether CABG is indicated should be considered carefully in younger children, and the

**Table 1** Indications of coronary artery bypass grafting for Kawasaki coronary disease [16]

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| Coronary artery bypass grafting (CABG) is indicated for patients with angiographically evident severe occlusive lesions of the coronary arteries and viability of myocardium in the affected area. Viability should be evaluated comprehensively, based on the presence/absence of angina and findings of ECG, thallium myocardial scintigraphy, two-dimensional echocardiography (regional wall movement), and other techniques |
| The following findings of coronary angiography are most important. When one of the following findings is present, consider surgical treatment |
| 1. Severe occlusive lesions in the left main trunk |
| 2. Severe occlusive lesions in multiple vessels |
| 3. Severe occlusive lesions in the proximal portion of the left anterior descending artery |
| 4. Jeopardized collaterals |
| In addition, the following conditions should also be considered in determining treatment strategy |
| 1. When the event is considered a second or third infarction due to the presence of chronic infarct lesions, surgery may be indicated. For example, surgery may be considered to treat lesions limited to the right coronary artery |
| 2. Lesions associated with recanalization of the occluded coronary artery or formation of collateral vessels should be evaluated especially carefully. Surgery may be considered for patients with findings of severe myocardial ischemia |
| 3. Whether CABG is indicated should be considered carefully in younger children based on long-term patency of grafts. In general, young children controllable with medical therapy are followed carefully with periodic coronary angiography to allow them to grow, while patients with severe findings have undergone surgery at 1–2 years of age. It is recommended that pedicle internal mammary artery grafts can be used in such cases as well |

decision should be based on long-term graft patency. In general, young children controllable with medical therapy are followed carefully with periodic coronary angiography (CAG) to allow growth, while those with severe disease undergo surgery at age 1–2 years.

In addition to the above guideline, CABG indications in Kawasaki coronary disease are also described in the American Heart Association (AHA) scientific statement [18]. Indications for coronary bypass procedures in children have yet to be established in clinical trials, but such surgery should be considered when reversible ischemia is present on stress-imaging test results, the myocardium to be perfused through the graft is still viable, and no appreciable lesions are present in the artery distal to the planned graft site (evidence level C). However, these indications are ambiguous and impractical in clinical situations.

The surgical indications for Kawasaki coronary disease in our center are listed in Table 2. In principle, coronary aneurysms should be carefully observed for 1–2 years under restrictive anticoagulation therapy, because coronary aneurysms regress in 50 % of patients within 1–2 years [17, 19]. The presence of a giant aneurysm ($\geq$8 mm in diameter) is a risk factor for thrombotic occlusion and progression of obstructive lesions [17, 20], and aneurysms $\geq$5 mm in diameter may become stenotic [21]. Giant aneurysms involving the LMT or both the LAD and RCA can cause fatal complications, whereas single-vessel obstruction of the RCA is often accompanied by marked development of collateral vessels without symptoms [22–24]. Single-vessel obstruction of the RCA is therefore not a basic surgical indication [23]. The presence of severe ischemia in giant coronary aneurysms involving obstructive lesions of the LMT or LAD is an unequivocal indicator for CABG at our center. Kitamura et al. limited their surgical indications to angiographically significant obstructive lesions (usually stenosis >75 %, and preferably >90 %), with ischemia [3, 5, 14]. In addition, giant aneurysms with recurrent thrombosis under restrictive anticoagulation therapy, or giant aneurysms with severely delayed flow without significant localized stenosis, are not indications for CABG, because they experienced early occlusion of the ITA graft due to competitive flow [14]. However, we consider that these conditions are indications for CABG when ischemia is obvious, because these conditions increase the risk of myocardial infarction. Use of a Doppler wire or pressure wire to measure average peak flow velocity (APV), coronary flow reserve (CFR), and myocardial functional flow reserve (FFRmyo) is useful in determining the functional severity of coronary artery stenosis and myocardial ischemia [25]. Nevertheless, surgical indications for giant aneurysm without significant stenosis should be considered carefully, because evaluating the degree of localized stenosis around giant aneurysms is difficult, owing to severe flow turbulence.

Early detection and treatment of myocardial ischemia is essential to prevent myocardial infarction and fatal complications. However, determining surgical indications for younger children is particularly difficult because of the technical difficulties involved. About one-third of KD patients who develop myocardial infarction have no obvious symptoms, probably because infants and younger children do not complain of chest pain [22]. Comprehensive evaluation of CABG candidates should comprise clinical signs and symptoms and findings from CAG, exercise electrocardiography (ECG), echocardiography, stress myocardial scintigraphy, left ventriculography (LVG), and other modalities [16]. In particular, pharmacological stress tests are useful for young children, who may not be able to tolerate exercise stress tests for evaluation of myocardial ischemia [26]. Surgical treatment is strongly recommended for children with a history of myocardial infarction and impaired left ventricular ejection fraction (LVEF), because about 60 % of survivors of first myocardial infarction have some degree of cardiac dysfunction [22], and impaired LVEF adversely affects prognosis [7]. In addition, patients who underwent surgical intervention soon after KD onset had fewer episodes of preoperative myocardial infarction and a lower incidence of postoperative cardiac events than did those who underwent surgical intervention later [27]. A recent report showed that CABG can be performed safely even in young children, in contrast to previous indications [3]. Application of plain old balloon angioplasty (POBA) for anastomotic stenosis of ITA grafts has improved graft patency in young children [28]. CABG may thus be indicated for severe ischemia in patients aged 1–2 years [16]. ITA grafts have also been used for congenital cardiac surgery in infants and young children [29, 30]. We believe that LITA-to-LAD grafting is the gold standard, even in children >1 year in age or >10 kg in body weight.

**Table 2** Surgical indications for Kawasaki coronary disease at our center

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|---|
| 1. Deliberate observation for 1–2 years, under restrictive anticoagulation therapy |
| 2. Surgical indications: positive ischemia with giant aneurysms involving the left main trunk or left anterior descending artery |
| 2.1 Stenotic or occluded coronary artery |
| 2.1 Giant aneurysm with recurrent thrombosis under restrictive anticoagulation therapy |
| 2.3 Giant aneurysm with severely delayed flow |

## *CABG at Our Center*

### Patients and Methods

Forty-one children and adolescents underwent CABG for Kawasaki coronary disease between 1991 and 2014 at Nippon Medical School Hospital in Japan, on the basis of our surgical indications. Mean age at surgery was $12.7 \pm 8.9$ years (range, 1–37 years); 32 (78 %) were male and nine (22 %) were female. There were 26 patients (63 %) aged $\leq$12 years and seven patients (17 %) aged $\geq$20 years. Mean age at KD onset was $3.5 \pm 2.8$ years (range, 4 months to 12 years; no data for 3 patients). Twelve patients (29 %) had a history of myocardial infarction and a preoperative LVEF of <35 % was noted in one patient (2 %). Four patients (10 %) had a history of percutaneous coronary intervention (PCI), including percutaneous transluminal coronary rotational ablation (PTCRA) in two patients (5 %). One patient with a history of PTCRA was eventually referred to our hospital after she had undergone PCI four times at another hospital. Two patients (5 %) had severe mitral regurgitation. The preoperative characteristics of the patients are summarized in Table 3.

Surgical indications at our center are listed in Table 2 and discussed in the previous section. Severe ischemia in a giant coronary aneurysm involving obstructive lesions of the LMT or LAD is an unequivocal indication for CABG. In addition, when obvious ischemia is detected, a giant aneurysm with recurrent thrombosis or severely delayed flow, without significant obstructive lesions, is an indication for CABG. Giant aneurysms were identified in 40 patients (98 %) by CAG. In the remaining patient without obvious coronary aneurysm, an occlusive lesion was found at the LMT. The aneurysms were located at the LMT in 17 patients (41 %), the LAD in 40 (98 %), the LCx in 16 (39 %), and the RCA in 38 patients (93 %) (Fig. 1). Most patients for whom CABG was indicated had aneurysms in both the LAD and RCA. Obstructive lesions developed at the inflow or outflow sites of coronary aneurysms in 31 patients (76 %) and at the LMT in three (7 %), LAD in 29 (71 %), LCx in six (15 %), and RCA in 21 patients (51 %). CABG was indicated for the remaining 10 patients (24 %) without obstructive lesions, because of giant aneurysms of the LMT or LAD with recurrent thrombosis or severely delayed flow. In all patients, we detected clinical signs of myocardial ischemia or positive signs of ischemia on ECG, echocardiography, or myocardial scintigraphy during exercise or pharmacological interventions.

**Table 3** Preoperative characteristics of patients

| | Value |
|---|---|
| Males | 32 (78 %) |
| Age at surgery, y (range) | 12.7 ± 8.9 (1–37) |
| Age ≤12 years | 26 (63 %) |
| Age ≥20 years | 7 (17 %) |
| Age at KD onset, y | 3.5 ± 2.8 |
| Prior myocardial infarction | 12 (29 %) |
| Low LVEF | 1 (2 %) |
| Previous PCI | 4 (10 %) |
| Severe MR | 2 (5 %) |

*KD* Kawasaki disease, *LVEF* left ventricular ejection fraction, *PCI* percutaneous coronary intervention, *MR* mitral regurgitation

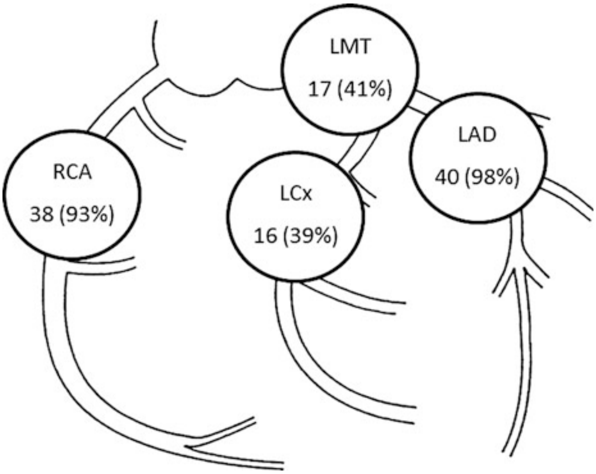


**Fig. 1** Distribution of coronary aneurysms in 41 patients undergoing CABG. Aneurysms were located at the LMT in 17 patients (41 %), LAD in 40 (98 %), LCx in 16 (39 %), and RCA in 38 patients (93 %)

## Operative Procedure

All patients underwent initial CABG. Thirty-nine (95 %) underwent conventional CABG under cardiopulmonary bypass and cardioplegic arrest. Among the remaining two patients, one (age 29 years) underwent on-pump beating CABG and the other (age 37 years) underwent off-pump CABG. Two patients (5 %) underwent concomitant mitral valve repair for coexisting severe mitral regurgitation. Nine patients (22 %) underwent concomitant down-sizing operation for giant aneurysms of non-LAD lesions.

The 41 patients received a mean of 1.6 ± 0.7 grafts: one graft for 20 patients (49 %), two grafts for 17 (41 %), and three grafts for four patients (10 %). Most patients (90 %) underwent revascularization of only one or two vessels. Forty patients (98 %) underwent ITA-to-LAD grafting (LITA in 37; RITA in 3) exclusively. The remaining patient, who had a history of successful PTCRA for the LAD, underwent two-vessel revascularization for the LCx and RCA, using the RITA and

**Table 4** Details of surgical procedures for CABG

| | |
|---|---|
| Type of CABG | |
| Conventional CABG | 39 (95 %) |
| On-pump beating CABG | 1 (2 %) |
| Off-pump CABG | 1 (2 %) |
| Concomitant procedure | |
| Mitral valve repair | 2 (5 %) |
| Down-sizing operation | 9 (22 %) |
| No. of distal anastomoses | 1.6 ± 0.7 |
| Single/double/triple | 20 (49 %)/17 (41 %)/4 (10 %) |
| Conduits used | |
| LITA/RITA/GEA | 40 (98 %)/14 (34 %)/3 (7 %) |
| ITA to LAD grafting | 40 (98 %) |
| Sequential grafting | 9 (22 %) |
| Y-composite graft | 2 (5 %) |
| Target coronary arteries | |
| LAD/Dx/LCx/RCA | 40/11/7/8 |

*CABG* coronary artery bypass grafting, *LITA* left internal thoracic artery, *RITA* right internal thoracic artery, *GEA* right gastroepiploic artery, *ITA* internal thoracic artery, *LAD* left anterior descending artery, *Dx* diagonal branch, *LCx* left circumflex artery, *RCA* right coronary artery

GEA to preserve the LITA for a forthcoming obstructive lesion of the LAD. All operations were completed using only arterial grafts, comprising the LITA in 40 patients (98 %), RITA in 14 (34 %), and GEA in three (7 %); no SVG was used. A total of 66 distal anastomoses were placed, for the LAD in 40, the diagonal branch (Dx) in 11, the LCx in seven, and the RCA in eight. Revascularization for the RCA was not completed in some patients in whom obstructive lesions in the RCA accompanied marked development of collateral vessels without ischemia. Sequential grafting, such as LITA-Dx-LAD, was performed in nine patients (22 %). A Y-composite graft was used in two patients (5 %). However, we avoided complicated graft designs, such as the Y-composite graft, since we have encountered graft occlusion due to a string phenomenon of the LITA graft in a Y-composite graft using bilateral ITA grafts. Details of the surgical procedure for CABG are summarized in Table 4.

## Results

There were no operative or hospital deaths, and no late deaths occurred during follow-up (mean duration, 10.9 ± 6.3 years). The cardiac event-free rate at 23 years after first-time CABG was 91.1 % (Fig. 2). Cardiac events occurred three times, in three patients. Two patients underwent redo CABG; the GEA as a free graft was anastomosed to the LAD at 1 year after primary surgery in one patient, because of occlusion of the LITA graft (used as a Y-composite graft) anastomosed to the LAD.

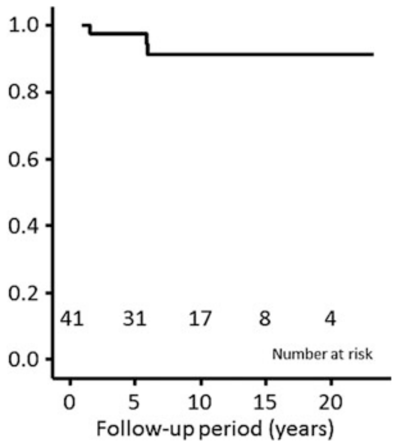


**Fig. 2** Cardiac event–free rate in the 41 patients. The cardiac event-free rate at 23 years postoperatively was 91.1 %. Cardiac events occurred in three patients, including redo CABG in two patients (for graft occlusion in one patient and for a new obstructive lesion in one patient) and ICD implantation for VT in one patient

The GEA as an in-situ graft was anastomosed to the RCA at 5 years after primary surgery in another patient, because of a new obstructive lesion. One patient received an implantable cardioverter-defibrillator (ICD) for ventricular tachycardia (VT) at 7 years after primary surgery. No patient suffered myocardial infarction postoperatively. All patients were symptom-free with no obvious restrictions in daily life.

Graft patency was evaluated postoperatively in all patients (by CAG in 40 patients; by multidetector-row computed tomography (MDCT) in one patient aged 37 years). Most patients underwent CAG postoperatively within 6 months after the primary operation, and about half underwent sequential CAG to evaluate mid- and long-term graft patency. The early graft patency rate was 98 % (65/66 anastomoses). The LITA graft (used as a Y-composite graft with the RITA) anastomosed to the LAD was occluded and required redo CABG at 1 year after primary operation, as described previously. No PCI was needed postoperatively.

## Case Presentations

Case 1 (Fig. 3)

A 1-year-old boy with a giant aneurysm of the LMT to the proximal LAD (with severely delayed flow) underwent CABG (LITA-LAD). The patency of the LITA graft was confirmed by CAG at 1 month postoperatively, and growth of the LITA graft was confirmed by CAG at 1 year postoperatively, suggesting that the ITA represented a “live conduit”.

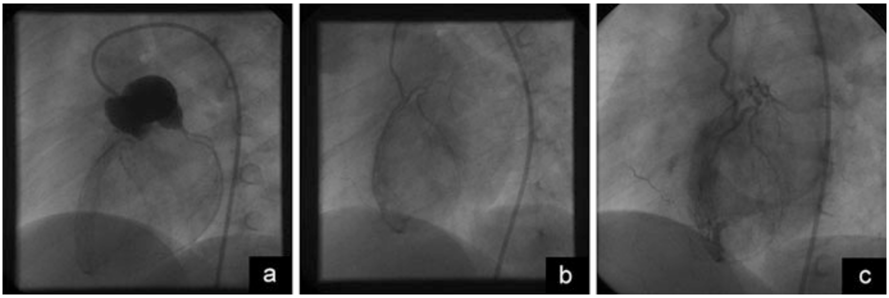


**Fig. 3** A 1-year-old boy underwent CABG (LITA-LAD) for a giant aneurysm of the LMT to proximal LAD, with severely delayed flow. (**a**) Preoperative CAG shows a giant aneurysm of the LMT to proximal LAD, with severely delayed flow. (**b**) Patency of the LITA graft is confirmed on CAG at 1 month postoperatively. (**c**) Growth of the LITA graft is confirmed on CAG at 1 year postoperatively

Case 2 (Fig. 4)

A 27-year-old man with giant aneurysms of the proximal LAD (with total occlusion) and proximal RCA (with segmental stenosis) and severe mitral regurgitation underwent CABG (LITA-Dx-LAD and RITA-RCA) and mitral valve annuloplasty. Patency of the LITA and RITA grafts and absence of mitral regurgitation were confirmed by CAG and LVG at 1 month postoperatively.

## Down-Sizing Operation for Giant Coronary Aneurysms

### *Introduction*

Giant aneurysms often develop at the proximal portion of the LAD and RCA [19, 22]. LITA-to-LAD grafting is the gold standard, whereas surgical revascularization for giant aneurysms of non-LAD lesions is controversial, particularly for young children, because of the lack of reliable grafts. Giant aneurysms impair coronary circulation with reduced APV, CFR and shear stress and a turbulent flow pattern [31–33]. This can induce thrombus formation inside aneurysms, which may predispose the patient to acute thrombosis, leading to myocardial ischemia and infarction.

Combination therapy using warfarin and aspirin reduces the risk of myocardial infarction in KD patients with giant aneurysms [34, 35], but several arguments against anticoagulation treatment have been raised, especially for young children. First, anticoagulation therapy in an infant obviously increases the risk of hemorrhagic complications, although a multicenter study revealed that hemorrhagic complications associated with combination therapy for KD patients were acceptably low, at 1.7 % per patient-year [34]. Second, the optimal duration of warfarin treatment has not been determined. Lifelong anticoagulation treatment is

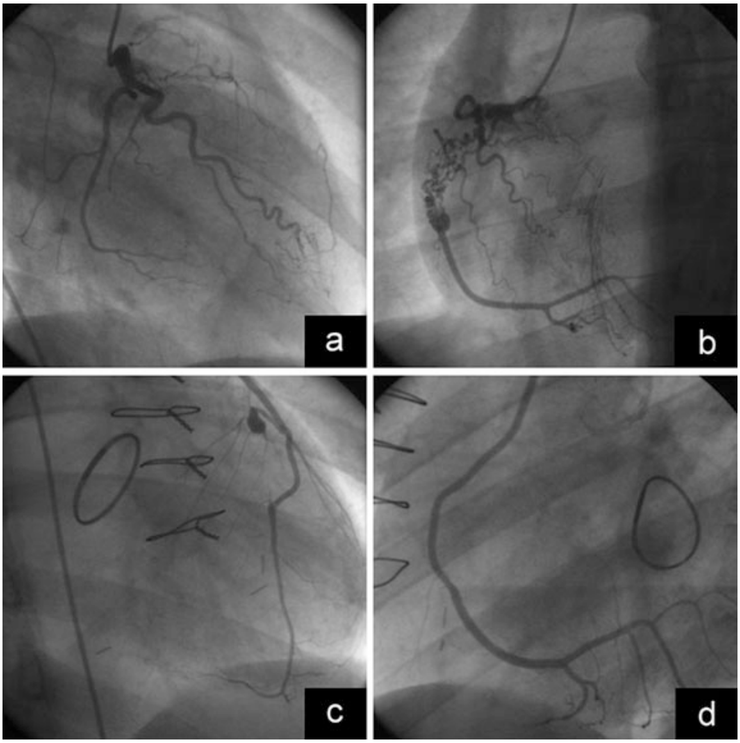


**Fig. 4** A 27-year-old man with giant aneurysms of the proximal LAD (with total occlusion) and proximal RCA (with segmental stenosis) and severe mitral regurgitation underwent CABG (LITA-Dx-LAD and RITA-RCA) and mitral valve annuloplasty. (**a** and **b**) Preoperative CAG shows giant aneurysms of the proximal LAD (with total occlusion) and proximal RCA (with segmental stenosis). (**c** and **d**) Postoperative CAG shows patency of the LITA and RITA grafts

recommended as long as giant aneurysms are present, unless regression of aneurysms is detected [34]. In addition, thrombus formation was observed in most giant aneurysms despite adequate anticoagulation [31].

The extent of flow disturbances associated with coronary artery aneurysms correlates with aneurysm size [31–33]. On the basis of this rheological finding, we have adopted a down-sizing operation for giant aneurysms of non-LAD lesions without stenosis and severe calcification, concomitant with CABG, to improve coronary circulation and allow discontinuation of warfarin.

## *Surgical Procedure of Down-Sizing Operation*

The procedure in the down-sizing operation for a giant aneurysm of the proximal RCA is shown in Fig. 5. After cardioplegic arrest, the giant aneurysm is exposed along its entire length, using an ultrasound scalpel, and incised longitudinally. Care is taken not to injure the right ventricular branches, to avoid perioperative right ventricular dysfunction. Fresh and old thrombi located on the posterior side of the distal aneurysm are carefully removed with a No. 15 knife and forceps. Interrupted mattress monofilament sutures are placed on the bottom of the aneurysm to make a “new” posterior wall for the RCA. The anterior wall of the “new” RCA is tailored with interrupted mattress monofilament sutures. A soft tube 5 mm in diameter is inserted into the internal lumen to maintain the appropriate size. Finally, the roof of

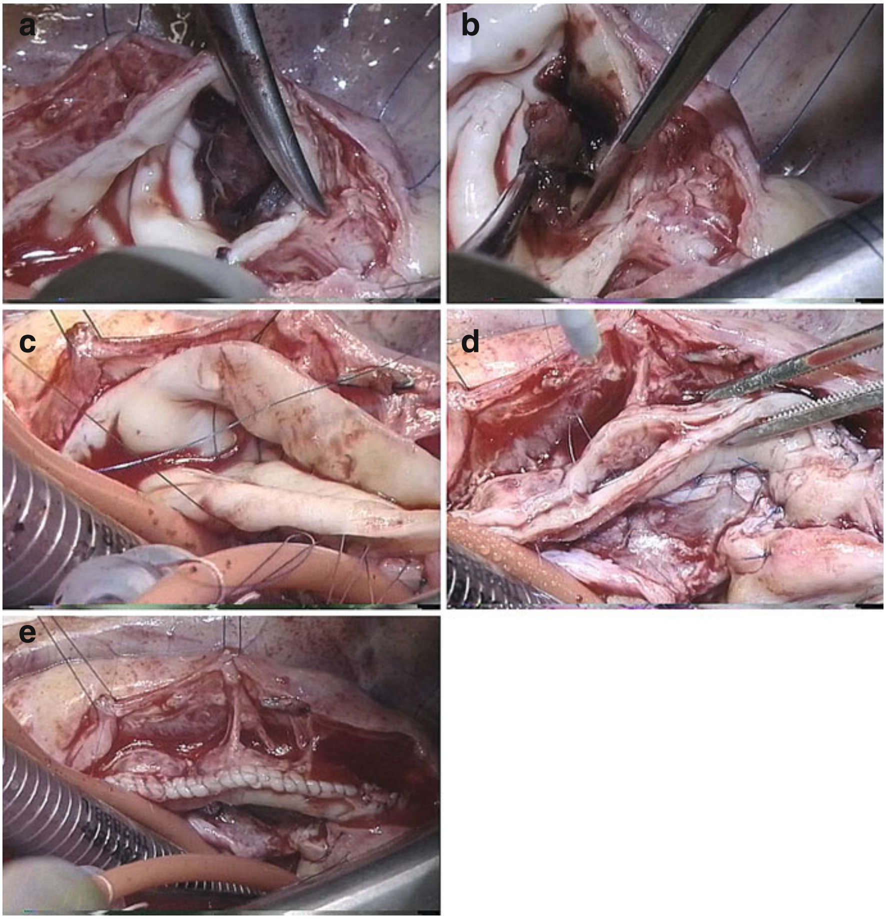


**Fig. 5** Surgical procedure in a down-sizing operation for giant aneurysm of the proximal RCA. (**a**) The giant aneurysm is exposed along its entire length and incised longitudinally to maintain an adequate distance to the ostium of the right ventricular branch. (**b**) Fresh and old thrombi located on the posterior side of the distal aneurysm are carefully removed using a No. 15 knife and forceps. (**c**) Interrupted mattress monofilament sutures are placed at the bottom of the aneurysm to form a new posterior wall for the RCA. (**d**) The anterior wall of the new RCA is tailored with interrupted mattress monofilament sutures, to reconstruct an internal lumen of appropriate size. (**e**) The roof of the RCA is closed by continuous monofilament sutures, to ensure hemostasis

the RCA is closed using continuous monofilament over-and-over sutures, to ensure hemostasis. Care should be taken to avoid stenosis around the transition between the aneurysm and "intact" RCA. To ensure an appropriate internal diameter around the transitional area, a small pericardial patch may be preferable.

## *Down-Sizing Operation at Our Center*

Nine patients underwent down-sizing operation for giant aneurysms of non-LAD lesions concomitant with conventional CABG. Mean age at surgery was $7.1 \pm 3.7$ years (range, 1–11 years). There were seven males (78 %) and two females (22 %). Mean age at KD onset was $4.4 \pm 2.9$ years (range, 1–9 years). Mean interval from KD onset to surgery was $3.2 \pm 3.6$ years (range, 4 months to 10 years). Giant aneurysms were located at the RCA in eight patients (proximal RCA in 7; distal RCA in 1) and at the proximal LCx in one patient. This procedure was applied mainly to sausage-like giant aneurysms in the proximal RCA. Surgical indications for a down-sizing operation were giant aneurysms without significant stenosis and severely calcified lesions. The degree of calcification was evaluated preoperatively by CAG and CT, and indication for down-sizing operation was decided based on operative findings (finger palpation). In addition, measurement of APV, CFR, and flow pattern by Doppler flow wire is essential to evaluate the functional severity of myocardial ischemia (as described in [25]). These parameters (APV $\geq$15 cm/s, CFR $\geq$2.0, and pulsatile flow pattern) were also measured postoperatively as an indicator of coronary circulation improvement and to evaluate the possibility of warfarin discontinuation.

The size of giant aneurysms was adequately reduced in five patients, who were able to discontinue warfarin due to improved parameters. Two patients continued warfarin because they could not complete the down-sizing operation sufficiently due to severe calcification. The intervals from KD onset to surgery in these two patients were 8 years and 10 years. Occlusion occurred in two patients. One patient underwent a down-sizing operation within 5 months after onset of KD. Occlusion was confirmed by CAG at 1 month postoperatively, without cardiac events. In the other patient, preoperative CFR was extremely low, indicating a disturbance of microcirculation. Patency of the RCA, with reduced size, was confirmed on CAG at 1 month postoperatively, but occlusion and collateral vessels were identified on CAG at 6 months postoperatively, without cardiac events. On the basis of these findings, we believe that conditions such as acute phase or existence of micro-circulatory disturbance may be contraindications for a down-sizing operation. The down-sizing operation for giant aneurysms of non-LAD lesions may be a good choice to improve coronary circulation and allow warfarin discontinuation, if suitable indications for this procedure can be established. Long-term patency of coronary arteries after a down-sizing operation should be assessed in the future study.

## *Case Presentation*

### Case 3 (Fig. 6)

An 11-year-old boy with giant aneurysms of the proximal LAD (with severely delayed flow) and the proximal RCA (without significant stenosis) underwent CABG (LITA-LAD) and down-sizing operation of the RCA. Postoperative CAG at 1 month postoperatively showed patency of the LITA graft and adequate

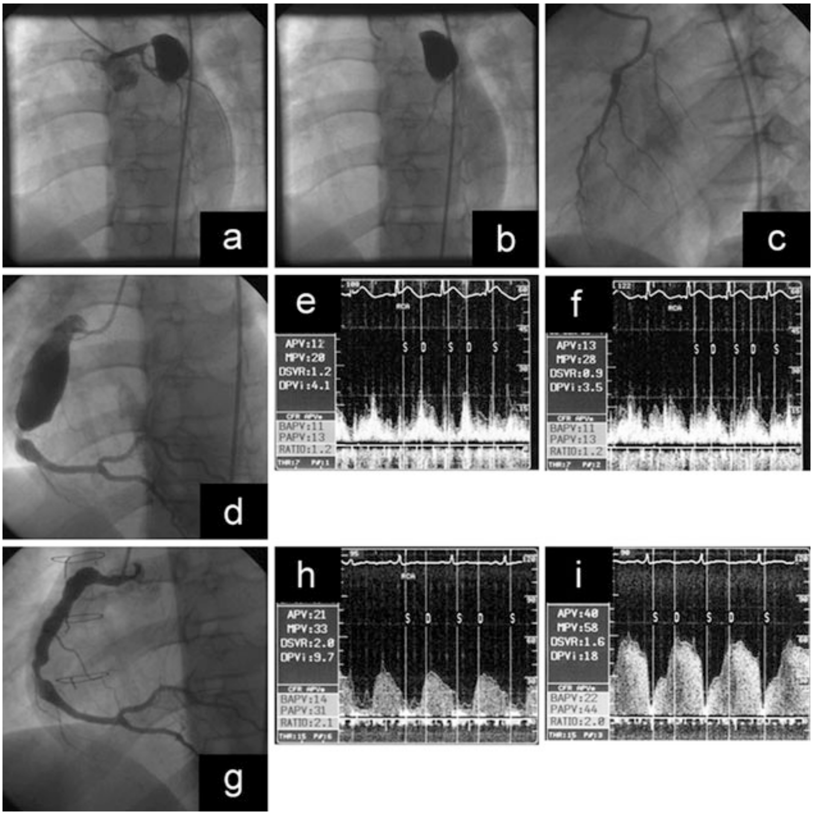


**Fig. 6** An 11-year-old boy underwent CABG (LITA-LAD) and down-sizing operation of the RCA for treatment of giant aneurysms of the proximal LAD (with severely delayed flow) and proximal RCA (without significant stenosis). (**a** and **b**) Preoperative CAG shows a giant aneurysm of the proximal LAD, with severely delayed flow (**a**: early phase; **b**: delayed phase). (**c**) Patency of the LITA graft is confirmed on CAG at 1 month postoperatively. Preoperative CAG shows giant aneurysm of the proximal RCA, without significant stenosis. (**e** and **f**) Coronary blood flow velocity is measured by Doppler flow wire. APV is 11 cm/s, CFR is 1.2, and the flow pattern is turbulent (**e**: at rest; **f**: after papaverine hydrochloride infusion). (**g**) Postoperative CAG at 1 month postoperatively shows that the size of the giant aneurysm of the RCA has been adequately reduced. (**h** and **i**) After down-sizing operation, APV is 21 cm/s, CFR is 2.0, and the flow pattern is pulsatile (**h**: at rest; **i**: after papaverine hydrochloride infusion)

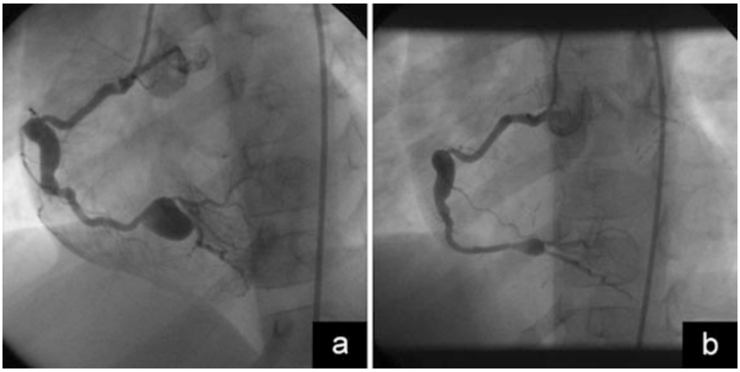


**Fig. 7** A 4-year-old boy with giant aneurysms of the proximal LAD (with severely delayed flow) and distal RCA (without significant stenosis) underwent CABG (LITA-LAD) and down-sizing operation of the giant aneurysm of the distal RCA. (**a**). Preoperative CAG shows a giant aneurysm of the distal RCA, without significant stenosis. (**b**). Postoperative CAG at 1 month postoperatively shows that the size of the giant aneurysm of the distal RCA has been adequately reduced

reduction in the size of the giant aneurysm of the RCA. Coronary blood flow velocity was measured by Doppler flow wire. Preoperatively, APV was 11 cm/s, CFR was 1.2, and the flow pattern was turbulent. After down-sizing operation, APV was 21 cm/s, CFR was 2.0, and the flow pattern was pulsatile. The patient was able to discontinue warfarin, due to these improved measurements. RCA patency was confirmed by CAG at 4 years postoperatively.

### Case 4 (Fig. 7)

A 4-year-old boy with giant aneurysms of the proximal LAD (with severely delayed flow) and distal RCA (without significant stenosis) underwent CABG (LITA-LAD) and down-sizing operation of the distal RCA. Postoperative CAG at 1 month postoperatively showed patency of the LITA graft and adequate reduction of the size of the giant aneurysm of the distal RCA. He was able to discontinue warfarin. RCA patency was confirmed by CAG at 1 year postoperatively.

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