

**Case
Report**

True Aneurysm of the Left Main Trunk in a Marfan Syndrome Patient at Remote Period after Bentall Operation and Total Arch Replacement: A Case Report

Kenshi Yoshimura, MD, Hideyuki Tanaka, MD, PhD, Tomoyuki Wada, MD, PhD, Takashi Shuto, MD, PhD, Madoka Kawano, MD, Hirofumi Anai, MD, PhD, and Shinji Miyamoto, MD, PhD

In 2002, a 37-year-old male with Marfan syndrome underwent the Bentall operation, total arch replacement, and aortobifemoral bypass for DeBakey type IIIb chronic aortic dissection, annuloaortic ectasia, and aortic regurgitation. In 2007, mild mitral regurgitation (MR) caused by mitral valve prolapse was identified. In April 2017, echocardiography revealed the worsening of MR and moderate tricuspid regurgitation (TR). Moreover, coronary angiography (CAG) revealed a coronary artery aneurysm in the left main trunk (LMT). In August 2017, the patient underwent mitral valve replacement (MVR), tricuspid annuloplasty (TAP), and coronary artery reconstruction. We reconstructed the LMT aneurysm using an artificial graft. True aneurysm of the coronary artery complicated with Marfan syndrome is a rare complication that has seldom been reported. This case highlights that it is essential to carefully follow-up patients with Marfan syndrome after the Bentall operation.

Keywords: coronary artery aneurysm, Marfan syndrome, Bentall operation

Introduction

In patients with Marfan syndrome, coronary artery anastomotic pseudoaneurysm following aortic root replacement had been reported as a complication. However, with the widespread use of the Carrel patch method, few cases of pseudoaneurysm have been reported. In contrast,

although true aneurysms of the coronary artery ostium have been reported, few cases of an aneurysm of the coronary artery itself exist. Surgical intervention is necessary to treat coronary artery aneurysms; however, the standard surgical procedure for such aneurysms is yet to be established. Herein, we report a case of a left main trunk (LMT) aneurysm accompanied by Marfan syndrome after the Bentall operation that could be reconstructed using an artificial graft.

Department of Cardiovascular Surgery, Oita University Hospital, Yufu, Oita, Japan

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Corresponding author: Kenshi Yoshimura, MD. Department of cardiovascular surgery, Oita University, 1-1 Idaigaoka, Hasama town, Yufu, Oita 879-5593, Japan
Email: k-yoshi@oita-u.ac.jp



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Case Report

In 2002, a 37-year-old male with Marfan syndrome underwent the modified Bentall operation, total arch replacement, and aortobilateral femoral artery bypass for DeBakey IIIb chronic aortic dissection, annuloaortic ectasia, and aortic regurgitation, in which aortic root and total arch replacements were performed using a quadricurcated graft with the elephant trunk technique. In addition to the replacement of the aortic valve with mechanical

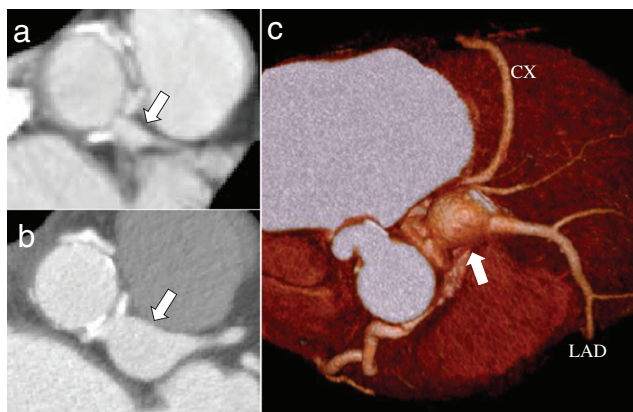


Fig. 1 (a) A postoperative contrast CT scan at initial surgery in 2002 revealed that there was no aneurysmal change in the ostium of the left coronary artery and the LMT (white arrow). (b) A contrast CT scan in 2017 revealed aneurysmal change in LMT (white arrow). (c) A coronary contrast CT scan (3D reconstruction) revealed that the LAD and CX were branched from the aneurysm (white arrow). CX: circumflex artery; LAD: left anterior descending artery; LMT: left main trunk

valve, the coronary arteries were reconstructed using the Carrel patch method, in which the coronary button was reinforced with Teflon strip at anastomosis. At the initial surgery, no aneurysmal change was observed in the LMT. In 2003, he underwent total thoracoabdominal aortic replacement surgery for the remaining chronic dissection. Postoperatively, he was regularly followed up as an outpatient. In 2007, he complained of chest pain during exercise along with fatigue of both the upper limbs. Computed tomography (CT) revealed no aneurysmal change in the coronary artery (**Fig. 1a**); moreover, echocardiography revealed normal left ventricular wall motion and only a mild mitral regurgitation (MR) due to mitral valve prolapse. Subsequently, he was conservatively treated as an outpatient for 10 years. Dilatation of LMT was detected during CT examinations conducted at annual follow-ups. In April 2017, echocardiography revealed MR exacerbation and moderate tricuspid regurgitation (TR). Coronary angiography (CAG) revealed no significant coronary stenosis but revealed a giant coronary artery aneurysm of the LMT, from which the left anterior descending (LAD) artery and circumflex (CX) artery were arising. Furthermore, coronary CT angiography clearly showed the aneurysm (**Figs. 1b and 1c**). The diameter of the aneurysm was 17 mm. No aneurysmal change was observed in the right coronary artery. Transesophageal echocardiography revealed severe MR and posterior cusp (P2-P3) prolapse. Besides, he also had moderate TR.

Accordingly, the patient underwent mitral valve replacement (MVR), tricuspid annuloplasty (TAP), and LMT reconstruction in August 2017. First, an 8-mm conduit was anastomosed to the side of the graft of the aortobilateral femoral artery bypass for the arterial return of the extracorporeal circulation (ECC). Subsequently, using median sternotomy, the adhesion was gently peeled to expose the ascending aortic graft, the brachiocephalic artery graft, and the aortobifemoral bypass graft, which were then secured. Next, as the first branch graft to the brachiocephalic artery was located close to the heart, we translocated it to the aortobilateral femoral artery bypass graft to secure a space to clamp the aorta and maintain the blood flow to the brachiocephalic artery after clamping the aorta (**Figs. 2a and 2b**). We established ECC with the arterial return cannula placed in the 8-mm conduit and inserted QuickDraw for venous drainage from the right femoral vein to the right atrium (**Fig. 2c**). Then, we clamped the aortic graft at the distal side of the transected first branch, through which cardioplegia solution was infused in both antegrade and retrograde manners to result in and maintain cardiac arrest. We made an incision on the right side of the left atrium and observed posterior mitral leaflet prolapse due to P2-P3 chordae rupture. The patient already had the mechanical aortic valve implanted, and the aortic root graft obstructed the surgical field-of-view to perform mitral valve repair. Moreover, he may have to undergo reoperation in the event of mitral valve failure at remote period after valve repair. MVR is considered safer and more secure than mitral valve repair. As a result, we performed MVR (ATS 31 mm; Medtronic, Minneapolis, MN, USA).

We transected the aortic graft at the proximal side of the clamping site and gently exteriorized the left coronary artery aneurysm. Teflon strip reinforcement was observed at the coronary button anastomosis site. We slipped the left coronary button anastomosis to peel to expose the aneurysm. The left coronary artery ostium was not dilated to be about 5 mm, and the dilatation of the LMT itself was observed. However, we anticipated difficulty in the anastomosis because the aneurysmal wall appeared thin and fragile and had collapsed in the state of cardiac arrest. We resected the ostium of the left coronary artery including the proximal half of the aneurysmal wall of the LMT. We placed eight mattress sutures on the periphery of the LMT just before the bifurcation using 5-0 Prolene with spaghetti and replaced the LMT with tube graft (J-graft 7 mm, Japan Lifeline, Tokyo, Japan) (**Fig. 2d**). The aneurysmal wall appeared extremely fragile and likely to easily tear following a shallow stitch.

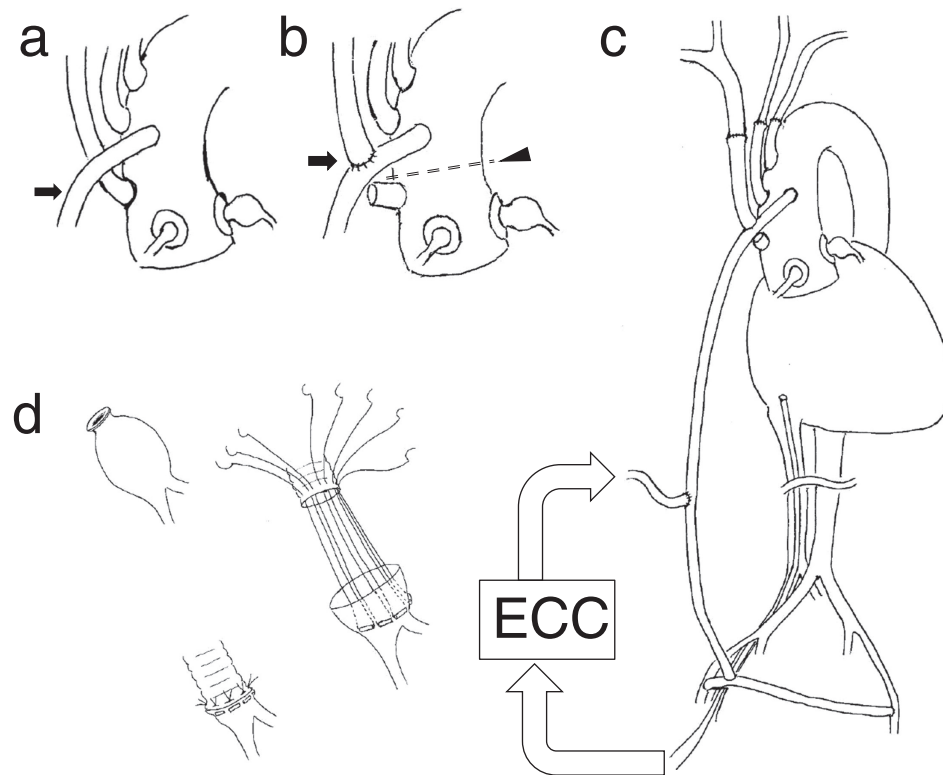


Fig. 2 (a) The first branch graft to the brachiocephalic artery was located close to the heart. There was aortofemoral bypass graft placed at the initial surgery on the ascending aortic graft (arrow). (b) The first branch graft was translocated to the aortofemoral bypass graft (arrow) to maintain the blood flow to the brachiocephalic artery after clamping the aorta (arrow head). (c) We established ECC with the cannula for the arterial return to the conduit on aortofemoral bypass graft and with the QuickDraw for venous drainage from the right femoral vein to the right atrium. (d) We replaced the LMT aneurysm with 7-mm artificial graft. Peripheral anastomosis was performed using eight mattress sutures. ECC: extracorporeal circulation; LMT: left main trunk

To reduce the aneurysm, we cautiously involved the distal half of the aneurysmal wall in the anastomosis to the maximum possible extent using deep stitches at the half-resected end. We completed LMT reconstruction by anastomosing the proximal side of the graft to the aorta. Then, we performed TAP (Physio tricuspid ring 28 mm; Edwards Lifesciences, Irvine, CA, USA). The aorta was declamped and the heartbeat restarted. Owing to the bleeding from the anastomotic portion of the coronary artery, we adjudged that hemostasis was difficult under pulsation; thus, the aorta was clamped again, and cardiac arrest was obtained by performing an antegrade cardioplegia. Although the anastomosis of the proximal side of the tube graft was removed to observe the distal anastomotic portion, there was difficulty in identifying the bleeding point. The entire circumferential reinforcement of peripheral anastomosis was made using eight stitches

of 4-0 Prolene. During re-anastomosis, we used deep stitches, which resulted in further reduction of the aneurysm. After declamping the aorta again, the patient's heartbeat spontaneously resumed, and the withdrawal of the ECC was smooth. However, the patient became liable to bleeding and restoring hemostasis became time-consuming. The total operation time was 17 h and 10 min, and the time for ECC was 11 h and 2 min.

The histopathological examination of the resected specimen, which comprised the wall from the ostium to the dilated coronary artery, revealed no atherosclerosis, calcification, inflammatory cell infiltration, and no evident degeneration, such as rupture of elastic fiber or cystic medial necrosis. The specimen wall had both elastic fibers and muscular parts. The ostium site comprised elastic fibers, and the distal site of the ostium, the dilated arterial wall, comprised muscular parts which was the

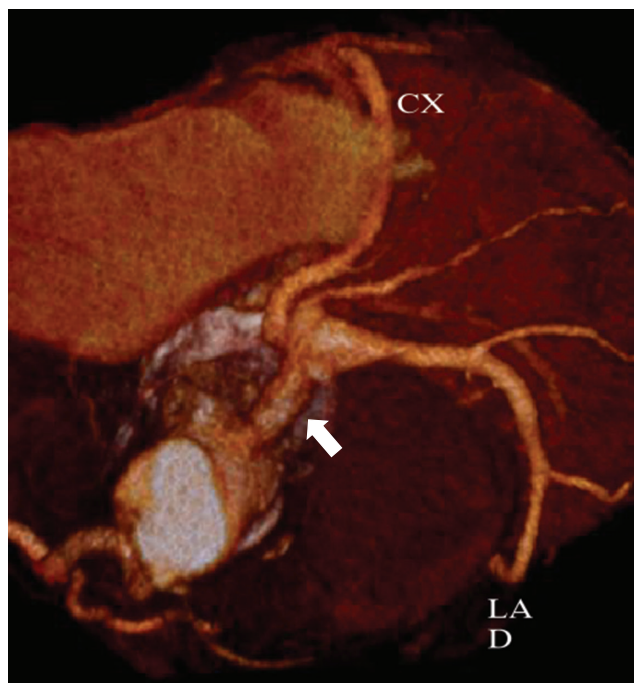


Fig. 3 A postoperative coronary contrast CT (3D reconstruction) revealed that the aneurysm was replaced with the artificial graft down just to the orifice of the LAD and CX (white arrow). CX: circumflex artery; LAD: left anterior descending artery

main component of the coronary artery. Histopathological and imaging findings exhibited no contradiction as the aneurysm originated from the wall of the coronary artery itself. The postoperative course was good. Furthermore, the left coronary artery aneurysm almost disappeared in the contrast CT (**Fig. 3**), and the blood flow of both LAD and CX was excellent. Although it seemed that the distal part of the aneurysm remained at the proximal LAD, the risk of rupture seemed low. However, follow-up with CT was considered necessary. On the 14th postoperative day, the patient was discharged.

Discussion

Anastomotic pseudoaneurysm has been a complication occurring in the anastomotic part of the coronary artery ostia after aortic root replacement surgery; however, with the introduction of the Carrel patch method, its frequency has decreased. In contrast, the number of cases of true coronary artery aneurysms increased after performing the Bentall operation. True aneurysms are categorized into two groups: one enlarging the aortic wall remaining in the anastomotic part of the coronary artery button and another dilating the coronary artery itself. Cases of the former type

are more frequently observed, and it is believed that the aortic wall remaining in the anastomotic part expands.¹⁻³⁾ Thus, in patients with Marfan syndrome, it is considered that the hole made in the graft should not be larger than the diameter of the respective coronary ostium to avoid the exposure of the diseased wall to circulating blood as far as possible. In addition, sutures used to anastomose the coronary buttons should pass through the rim of the ostium rather than through the surrounding aortic wall.²⁾

Conversely, aneurysms of the coronary arteries rather than the coronary ostium have been reported.⁴⁻⁷⁾ In our case, no aneurysmal change was noted in the anastomotic portion of the coronary artery, and no dilatation of the coronary artery ostium was observed. Besides, the LAD and CX arose from the aneurysm individually. Thus, we considered that the coronary artery itself must be an aneurysm in this case; this complication is rare and only three such cases⁴⁻⁶⁾ have been reported to date. In all aforementioned studies, including ours, the patients had Marfan syndrome. Although no histological changes were observed in the present case, there is a high possibility that such changes were caused by the histological degeneration rather than a technical problem in anastomosis because the LMT itself became aneurysmal.

Regarding the surgical procedure, each report explained a different mode of coronary artery reconstruction, and it is apparent that the standard surgical method has not been established yet. Onoda et al.⁴⁾ reported a case of coronary artery aneurysm after the original Bentall operation and performed distal anastomosis for coronary artery bypass grafting (CABG) using saphenous vein grafts (SVGs) and over sewed the stump of the LMT just at the LAD and CX. Okamoto et al.⁵⁾ reported a case of a giant left coronary artery true aneurysm after performing the modified Bentall operation and reconstructed the LMT by interposing the Dacron graft. Oyama et al.⁶⁾ reported a case of both left and right coronary artery true aneurysms following the Bentall operation, in which they reconstructed both the coronary arteries by replacement with great SVGs. In our case, as the coronary artery aneurysm extended not only to the LMT but also to the LAD, CX was in a state of being branched from the coronary artery aneurysm. When adopting the method of revascularization using CABG, two branch bypasses of the LAD and CX were considered necessary. However, adhesion because of previous operations was so intense that the approach to the CX was deemed to be challenging. Moreover, we believed that CABG with SVG, which had a concern about graft occlusion at a remote period, should be avoided for young patients.

Hence, we adopted the method of reconstruction using an artificial graft by resecting the aneurysm just before the LAD and CX branching.

As we could not obtain good surgical fields, bleeding was observed from the anastomotic site after the weaning of the ECC. A case report mentioned that the transection of the pulmonary artery facilitated the creation of a favorable surgical field for the repair of a false aneurysm in the LMT⁸; perhaps, this approach could also have deemed useful in our case.

Reportedly, coronary aneurysms may not only cause myocardial infarction⁷ due to thrombosis and embolism but also pose a risk of rupture,⁹ which is considered to be adapted for surgery when it is 3–4 times larger than the original coronary artery.^{10,11} Hence, in patients with Marfan syndrome who undergo aortic root replacement, following up using imaging by periodic CT is imperative.

Conclusion

True aneurysm of the coronary artery after performing the Bentall operation is extremely rare. For such cases, a repair technique has not yet been established. In determining the surgical procedure, it is necessary to consider factors such as the patient's age, state of adhesion, and site of lesion. In the present study, the lesion was successfully reconstructed using an artificial graft.

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Disclosure Statement

The authors declare no conflicts of interest associated with this manuscript.

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