



Case Report

Reversible left ventricular dysfunction due to severe stenosis of the elephant trunk graft: A case report



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ABSTRACT

Acquired coarctation of the aorta (CoA) following total aortic arch replacement (TAR) is a rare complication inducing left ventricular (LV) dysfunction probably due to increased LV afterload and secondary hypertension caused by increased upper body and decreased renal blood flow. We describe a case of a 35-year-old male who developed atypical CoA with severe LV dysfunction with LV ejection fraction of 10%, but without secondary hypertension after TAR using conventional elephant trunk (ET) technique for acute aortic dissection. Computed tomography revealed near-occlusive CoA due to narrowed distal ET. Because the myocardial histological findings were mild, and he had no cardiac failure history, we determined that LV function might be reversible. He underwent thoracic endovascular aortic repair (TEVAR), resulting in restored LV function. However, as the descending aortic false lumen distally to the end of ET was rapidly dilated, probably due to increased cardiac output and lower body blood flow, he underwent descending aortic replacement 3 months after TEVAR. In conclusion, a narrowed distal ET may cause LV dysfunction early after TAR, even without secondary hypertension. TEVAR may be a useful therapeutic option for a narrowed distant ET but can induce distal aortic dilatation.

<Learning objective: A 35-year-old male demonstrated that acquired coarctation of the aorta due to a narrowed distant elephant trunk led to left ventricular (LV) dysfunction with comparable severity of dilated cardiomyopathy even with normal blood pressure. Thoracic endovascular aortic repair is an effective treatment option to restore LV function by decreasing LV afterload. However, a narrowed distal ET relief with an increased cardiac output might cause distal aortic dilatation.>

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Introduction

Congenital coarctation of the aorta (CoA) has been reported to reduce left ventricular (LV) function [1,2], probably due to increased LV afterload and secondary hypertension caused by increased upper body blood flow and decreased renal flow. Acquired CoA with LV dysfunction is much rarer than congenital CoA but

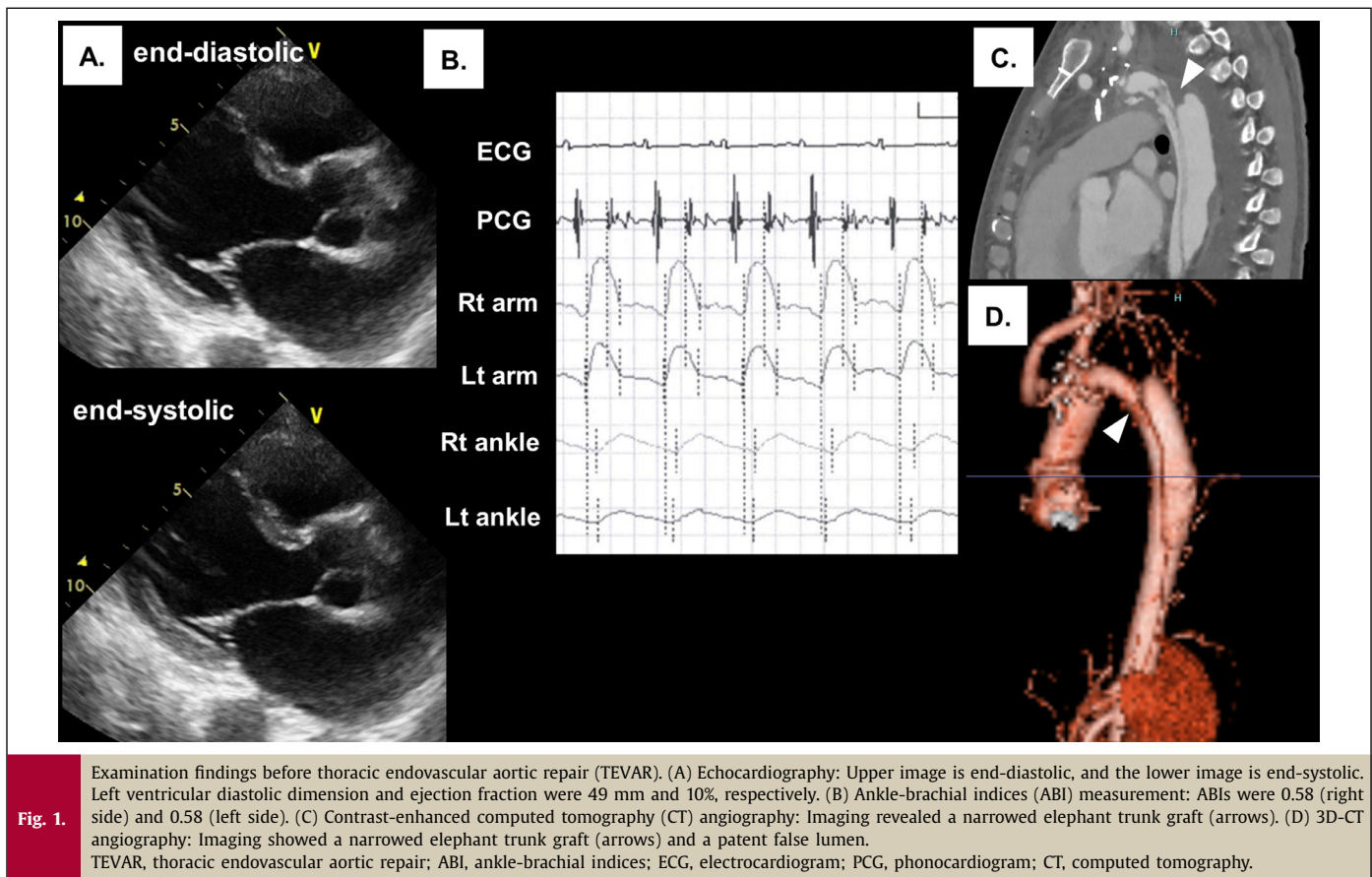
has been reported post-trauma [3,4] as well as post-aortic surgery [3,5]. Herein, we report a patient with severe LV dysfunction after total aortic arch replacement (TAR) with conventional elephant trunk (ET) technique for Stanford type A acute aortic dissection (AAAD).

Case report

A 35-year-old Japanese male was hospitalized at another hospital with sudden onset chest pain. He had a brother who died of

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AAAD at the age of 35 years. His blood pressures (BPs) were 96/40 and 66/30 mmHg in the left and right arm, respectively, and the lower limbs were pale. Contrast-enhanced computed tomography (CT) revealed AAAD extending from the ascending aorta to common iliac arteries with a patent false lumen. A reentry was observed at the renal artery level. He underwent emergent TAR with a 4 branched J Graft® (24 mm; Japan Lifeline Co., Ltd, Tokyo, Japan) using ET technique with a straight J Graft® (20 mm) through a median sternotomy. An aortic pathological examination showed severe medial degeneration with multifocal mucoid extracellular matrix accumulation, elastic fiber fragmentation, and smooth muscle cell nuclei loss.

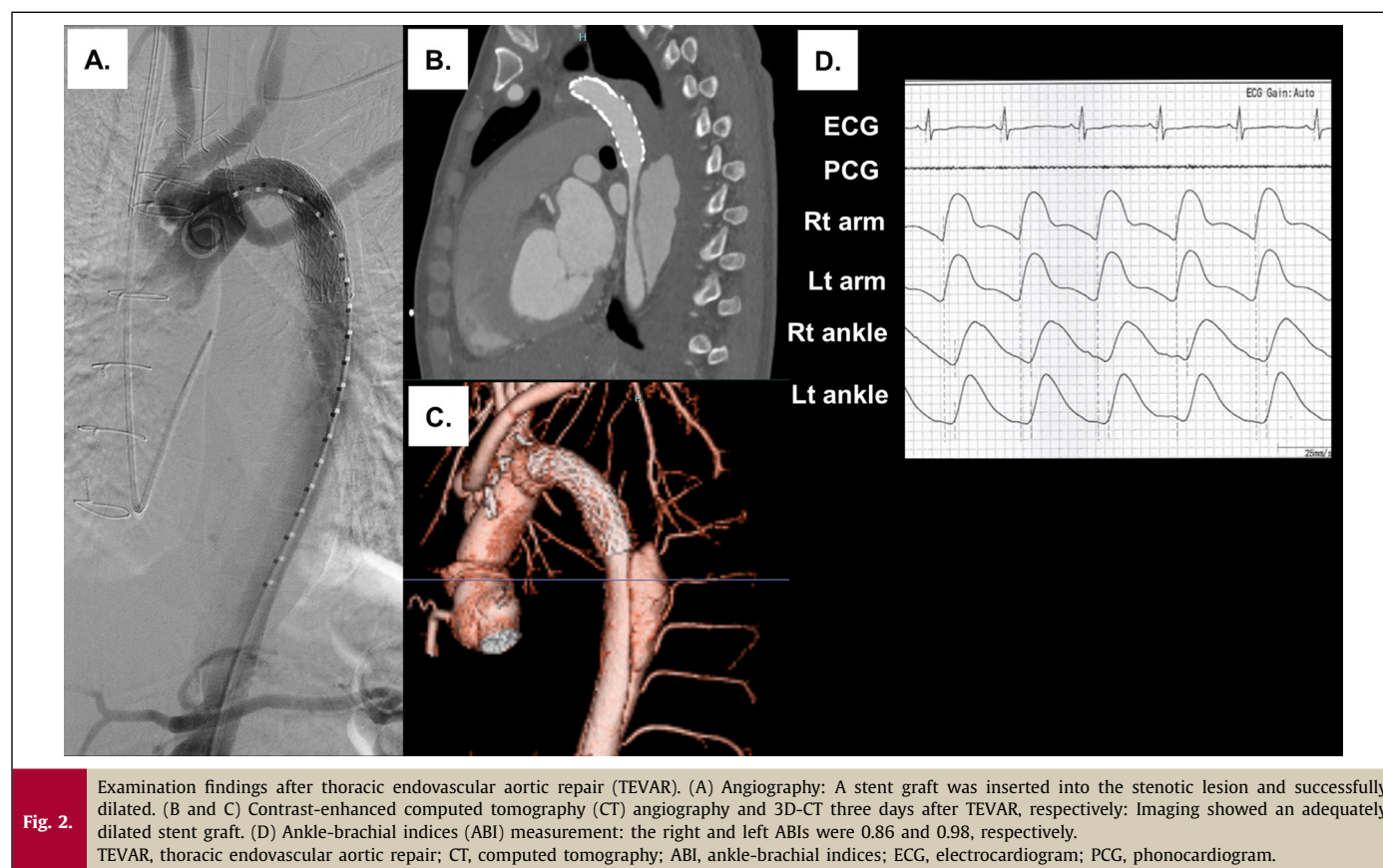
On the 10th postoperative day (POD), BP was 106/48 mmHg at the right arm, and the LV function was normal. He had the lower limb's compartment syndrome and underwent continuous renal replacement therapy (CRRT) due to acute kidney injury. After withdrawal from CRRT, he developed congestive heart failure. On the 32nd POD, echocardiography revealed LV ejection fraction (LVEF) of 30%. He had no hypertension during this period. As the coronary CT angiography revealed no coronary artery disease and the endomyocardial biopsy showed mild fibrosis without any secondary cardiomyopathy findings, dilated cardiomyopathy was suspected. LVEF further decreased to 10% (Fig. 1A). He became dependent on continuous inotrope infusion and was transferred to our hospital for advanced therapy, including mechanical circulatory support on the 83rd POD.

On admission, BPs were 60/40 mmHg and 100/60 mmHg at the lower and upper extremities (Fig. 1B). A systolic murmur was heard over the second left sternal border with radiofemoral delay. Right heart catheterization (RHC) revealed a pulmonary arterial

wedge pressure (PAWP) of 21 mmHg and a cardiac index (CI) of 2.4 L/min/m². CT angiography revealed a narrowed distal ET graft (Fig. 1C, D). The myocardial histological findings were mild, and he had no cardiac failure history; we speculated LV dysfunction might be reversible by LV unloading.

He underwent thoracic endovascular aortic repair (TEVAR) for ET graft expansion on the 85th POD. After systemic heparinization, direct pressure measurement revealed 78/53 mmHg and 108/52 mmHg at the abdominal aorta and the left radial artery, respectively. After confirming the dilation of stenotic portion by the inflation of balloon catheter, the GORE® TAG® Conformable Thoracic Stent Graft (TGU212110J; W.L. Gore & Associates, Inc., Flagstaff, AZ, USA) in both proximal and distal diameters to 21 mm was deployed from just below the reconstructed branch of the left subclavian artery to the true lumen of the middle descending thoracic aorta, to expand the ET graft stenosis. Post-TEVAR angiography and catheterization confirmed no endoleak (Fig. 2A–C), and there was a relief of upper-to-lower-extremity systolic blood pressure (ULSBP) gradient (Fig. 2D). Examination two months post-TEVAR revealed improved LV function (PAWP 3 mmHg and CI 3.1 L/min/m² in RHC and LVEF 32% in echocardiography).

Three months post-TEVAR, LVEF increased to 46% with no symptoms. However, the follow-up CT revealed a rapid expansion of the descending aortic false lumen from 35 × 30 mm at TEVAR to 43 × 36 mm (Fig. 3A, B). He underwent descending aortic replacement. The graft was anastomosed proximally to the distal end of ET graft and the native aortic wall together and distally to a double-barrel stump of the abdominal aorta (Fig. 3C). The postoperative course was uneventful, and currently, at 12-month follow-



up visits, he was in good physical condition with LVEF of 60% and BP of 94/57 mmHg.

Discussion

This case raised three important issues. First, LV dysfunction occurred from a narrowed ET graft even with well-controlled blood pressure, which is reversible by releasing the causative graft stenosis early post-TAR. Second, TEVAR might be a feasible therapeutic option for a narrowed ET graft, even in patients with LV dysfunction. Third, the relief of a narrowed ET graft with an increased cardiac output may cause distal aortic dilatation.

Previous studies reported that increased LV afterload and secondary hypertension caused by increased upper body blood flow and decreased renal blood flow play a role in causing LV dysfunction in acquired CoA [2–4] as well as congenital CoA [6,7]. However, endograft collapse may occur after TAR using ET as well as TEVAR [8,9]. To our knowledge, LV dysfunction has not been reported early after TAR. In TEVAR registries [8,9], 60% of endograft collapse events were noted in implants for aortic trauma. Approximately 30–40% of the endograft collapse patients had symptoms, such as hypertension, claudication, and acute renal failure, but not LV dysfunction, probably because the period from initial TEVAR to diagnosis might not be long enough to induce LV dysfunction.

On the other hand, Keen and Johnson [3] reported an 18-year-old male with acute LV dysfunction (LVEF 20%) due to endograft collapse (90%) caused by blunt trauma to his abdomen with a basketball one month after TEVAR for post-trauma CoA. The ULSBP gradient was 100 mmHg. Emergent new cuff graft placement restored LV dysfunction (LVEF 55%) and upper body hypertension soon after surgery. This case suggested that sudden and high-grade aortic narrowing by trauma could cause uncontrollable hyperten-

sion and LV dysfunction even soon after TEVAR endograft collapse. Much earlier ULSBP measurement than in the present case might be valuable for early diagnosis and therapeutic strategy decision to avoid LV dysfunction.

It is unclear why this patient showed severely reduced LV function despite a lack of secondary hypertension, which is an important afterload component. Prolonged bed rest due to several complications post-TAR might decrease organ blood flow demand, and oral administration of enalapril in a dose of 5 mg/day might have suppressed an enhancement of the renin-angiotensin system and subsequent secondary hypertension. Plasma aldosterone level on transfer in the present case remained within the normal range (246 pg/mL). Concerning secondary hypertension, Egbe et al. [1] reported that 220 of 546 congenital CoA adult patients were normotensive before therapeutic intervention. In this study, Doppler CoA pressure gradient, systolic blood pressure, or ULSBP gradient did not have a significant correlation with LV diastolic function indices or exertional symptoms. In contrast, the aortic isthmus ratio had the strongest correlation with them. Therefore, these data suggested that hypertension is not an essential factor to lead to LV dysfunction. Elevated arterial stiffening leading to lowered passive arterial expansion and subsequent counter-pulse wave elevation is another important afterload component [10]. The sudden onset anatomical stenotic changes from ET might have led to extremely elevated central artery stiffening, resulting in an elevated afterload against the LV. As tachycardia was observed at around heart rate of 100 bpm in the postoperative acute phase, excessive hyperactivity of the sympathetic nervous system might have elevated LV afterload. These factors may be the reason for LV dysfunction. However, there were no objective data, such as serum noradrenaline level. Therefore, we could not show the exact an-

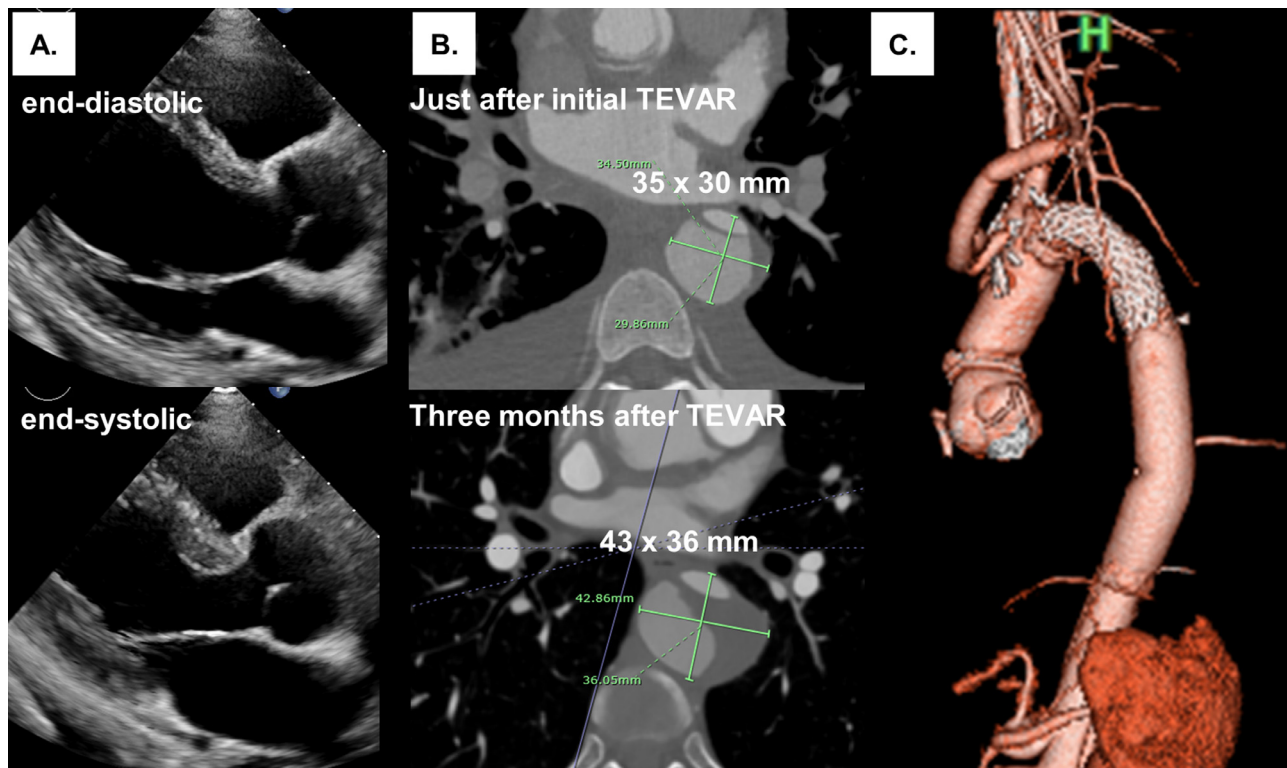


Fig. 3.

Examination findings before and after the second surgery (descending aortic graft replacement). (A) Echocardiography: Upper and lower images were taken at end-diastolic and end-systolic 3 months after thoracic endovascular aortic repair (TEVAR). Left ventricular diastolic dimension and ejection fraction were 44 mm and 46%, respectively. (B) Contrast-enhanced computed tomography (CT) angiography: The upper and lower images were taken just after and 3 months after TEVAR, respectively. The aorta size was 35 × 30 and 43 × 36 mm just after and three months after TEVAR, respectively. Images revealed an expanded pseudo-lumen of the descending aorta. (C) 3D-CT after descending aortic graft replacement: The prosthetic graft's proximal end was anastomosed to the distal end of the elephant trunk graft and native aortic wall together. TEVAR, thoracic endovascular aortic repair; CT, computed tomography.

answer to this important question of why LV function was severely reduced in the present case.

After the collapsed graft diagnosis, most patients undergo a reintervention [1,8,9]. Although a reentry was observed at the renal artery level in the present case, a less invasive strategy was selected because he had LV dysfunction. A narrowed distal ET graft was expanded, and the endograft was deployed from the distal end of ET graft to the true lumen of the descending thoracic aorta. This method diminished the ULSBP gradient and restored LV function. However, the descending aortic false lumen was rapidly dilated because of increased lower body blood flow and suspected aortic connective tissue disorder.

In conclusion, we encountered an atypical acquired CoA patient with severe LV dysfunction early after TAR using ET technique. Clinicians should know that unexplained LV dysfunction occurs from a conventional elephant trunk graft stenosis, even without secondary hypertension. ULSBP measurement may be valuable to evaluate the cause of reduced LV function even in this situation. TEVAR is a useful therapeutic option for treating ET graft stenosis. However, it should be noted that the relief of a narrowed distal ET graft with an increased cardiac output might cause distal aortic dilatation.

Declaration of Competing Interest

The authors declare that there is no conflict of interest.

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