Title Page

Successful Deployment of an Iliac Limb Graft to Repair Acute Aortic Rupture after Balloon Aortoplasty of Recoarctation in a Child with Turner Syndrome

I-Hui Wu[1], M.D, Mei-Hwan Wu[2], M.D. Shy-Jye Chen[3] M.D. Chung-I Chang[1], M.D.

[1]Department of Surgery, Cardiovascular Division, National Taiwan University Hospital

[2]Department of Pediatrics, National Taiwan University Hospital

[3]Department of Medical Imaging, National Taiwan University Hospital

Address for reprints: Chung-I Chang, M.D., Department of Surgery, Cardiovascular section, National Taiwan University Hospital, No 7, Chung-Shan S. Road, Taipei, Taiwan (Tel: 886-2-23123456 ext 65072/ FAX: 886-2-23225697 / E-mail address: joey@ntuh.gov.tw)

Word Counts: 1458 words

Article type: Case Report

Abstract

Aortic rupture is a rare but potentially catastrophic complication following balloon aortoplasty for recoarctation. We presented a 9 year-old patient with Turner syndrome, who experienced acute aortic rupture after balloon aortoplasty for recoartaction, was successfully rescued by antegrade deployment of a commercial available iliac limb extension stent-graft via ascending aortic conduit. Possible reasons for aortic rupture in this case included oversized ballooning based on angiographic measurement only, the inherent aortopathy and previous surgical repair. In agreement with earlier reports, the case we describe here also confirms the importance of keeping the commercially available stent graft available to treat the complication.

Introduction

The incidence of recurrent coarctation(re-CoA) after the surgical repair in neonate stage ranges from 5% to 50% in different reports[1]. Conventional management of re-CoA has been akin to the surgical treatment. However, post-surgical complication after re-do surgery carries significant mortality and morbidity. Treatment of CoA with a balloon catheter was first described in 1982 by Singer and colleagues[2], and had been an alternative method of managing re-CoA. This is certainly a less invasive method of intervention but well-described complications may occur. Aortic rupture is a rare but potentially catastrophic complication following balloon dilatation of re-CoA. Here we report a pediatric patient with Turner syndrome, who experienced acute aortic rupture after balloon aortoplasty for re-CoA, was successfully rescued with a commercially available self-expandable iliac limb stent graft.

Case Report

This was a 9-year-old girl with a history of Turner syndrome, juxtaductal CoA status post patch aortoplasty and patenet ductus arteriosus ligation through left thoracotomy at the age of one month old. After the age of eight, she was noted to have upper extremity hypertension and couple episodes of seizures. Follow-up doppler echocardiographic study and computed tomography (CT) revealed increasing

pressure gradient across the aortic isthmus and a discrete re-CoA distal to the left subclavian artery. The patient then underwent cardiac catheterization with a view to balloon aortoplasty of her re-CoA. The procedure was performed under local anesthesia, during which the patient also received 2,500 units of heparin intravenously. Angiographic diameter measurements of the proximal aortic arch, re-CoA, and distal descending aorta were 12.2, 7.8, and 13.9mm, respectively (Figure 1A). A pull-through pressure gradient of 30 mm-Hg between aortic arch and proximal descending aorta was documented. A 14mmx30mm Tyshak II angioplasty balloon (NuMED Inc., Hopkinton, NY, USA) was then advanced to the CoA site from the right groin. We gently inflate the balloon to 5 ATM to dilate the stenosis (Figure 1B). After ballooning, this patient complained of severe back pain. This was followed by a rapid fall in the patient's systemic blood pressure after the balloon was deflated. Immediate aortography disclosed extravasation of contrast medium to posterior aspects of the aorta. (Figure 1C). The balloon was advanced back and reinflated to tamponade the aortic rupture. Heparin was reversed with protamine while plasma expanders were administered to stabilize her hemodynamics. Because of the high risk of emergent re-do open thoracotomy repair, endovascular repair was discussed with her parents and planned. During perioperative angiogram, non-flow limited right common iliac artery dissection was founded (Figure 1D). Due to her small and

dissected iliac access, an eight mm Dacron graft was sewn to the ascending aorta via sternotomy as an antegrade conduit to put in a commercially available self-expandable stent graft. Because of her small diameter of thoracic aorta, we had to choose the off-label use of 16mm diameter iliac leg extension of the abdomimal aortic stent graft (ELSE-16-55, Cook, Bloomington, Indiana) to cover the ruptured aortic lesion. The stent graft was carefully positioned and sealed the aortic rupture. The left subclavian artery (LSA) was covered but retrogradely filled from the left vertebral artery, as confirmed by postoperative aortography (Figure 2A). The patient was then transferred to the intensive care unit and subsequently extubated with an uncomplicated recovery. Follow-up contrast CT at 7 days postoperatively confirmed no residual re-CoA, endoleak, or pseudoaneurysm formation (Figure 2B and C). The patient was discharged 14 days after the operation in good condition and was directed to take aspirin. In 6-month follow up, the patient had no symptoms of left arm weakness or stroke. The echocardiography showed no residual CoA.

Comment

Patients with Turner syndrome have a higher incidence of CoA, with a prevalence ranging from 4-14%[3]. The traditional management of CoA in these patients has been surgical repair. Less invasive balloon aortoplasy was first described in 1982 by Singer and colleagues[2]. Though limited data exist on the results of

balloon angioplasty for native CoA or re-CoA in patients with Turner syndrome, this procedure has been recommended as an acceptable alternative to surgical intervention by more encouraging early and mid-term results. Balloon aortoplasty became the first option of treatment for re-CoA in our institution after 2000. Although good results have been reported, this technique is not without sequelae, such as aortic rupture, dissection of the aorta; and vascular complications associated with the femoral approach itself (1-2%)[4]. The patient described here had the most life-threatening complication that could occur—aortic rupture—which was detected in its early stages and successfully rescued with the stent graft.

The presence of aortic cystic medial necrosis has been documented in patients with Turner syndrome. In addition to the intrinsic defect of the aorta, balloon oversizing and the formation of periaortic fibrous scarring subsequent to surgically repaired CoA are all possible etiologies for acute aortic rupture in our patient[5]. In Ozawa et al.[6] report, the chosen balloon size should not exceed the size of the healthy aorta at the level diaphragm, and the balloon-coarctation ratio should preferably not exceed 1.3:1. In our patient, the choice of balloon diameter based on angiographic measurement only is also a possible reason for oversizing the balloon. Report from Hijazi et al[7] has shown the coarctated segment should be measured carefully with methods such as magnetic resonance imaging, in addition to

angiography. When discrepancies are found between angiographic and MRI findings, intravascular ultrasound appears to be a promising and reliable means.

In adults, studies have shown in selective patients, LSA coverage can be accomplished safely in emergency settings and without revascularization. Staged LSA revascularization may be necessary but can be performed without long-term detriment to the left arm[8]. However, limited data are available for pediatric patients. In our patient, after 6 month follow, she develops no neurological or left arm symptoms. We plan to follow up this situation. The incidental finding of non-flow limiting right common iliac artery dissection may result from puncture of the inherent diseased aorta and the urgent advancement of tamponade balloon for acute aortic rupture. In our case, we chose a sternotomy approach for the stent graft deployment due to the small iliac access and the iatrogenic right femoral artery dissection. The off-label use of iliac limb extension stent graft is because of the size limitation of the commercially available thoracic stent graft, and also the unavailablity of commercial balloon expandable covered stents in our country..

In summary, what presented here is the successful management of aortic rupture after balloon aortoplasty of re-CoA with patch aortoplasty in a child with Turner syndrome. Caution should be advised for intervention in Turner syndrome patients with re-CoA due to the inherent aortic pathology and previous surgical repair.

Meticulous femoral puncture, sheath introduction, gently ballooning and the avoidance of oversizing the balloon are keys to avoid catastrophic complications. In common with earlier reports[5], the case that we describe here confirms the importance of keeping either self-expandable or balloon-expandable stent grafts available to cope with complications that have potentially fatal consequences in the event of emergency.

Figure Legends

Figure 1: (A): Aortography shows a discrete re-CoA (Arrow) distal to the left subclavian artery. The angiographic diameter measurements of distal aortic arch (white dashed line), re-CoA, and distal descending aorta (black dashed line) are 12.2 ,7.8, and 13.9 mm respectively

(B) After ballooning to 5 ATM, there is still a minimal waist in the balloon. (Arrow)

(C) Contrast extravasation of the ruptured aorta in post-dilatational aortography (Arrow)

(D) Iatrogenic retrograde right iliac artery dissection with patent flow to the right common femoral artery without contrast extravasation (Arrow)

Figure 2: (A) Endovascular stent graft deployment with complete seal of aortic rupture and retrograde filling of left subclavian artery from vertebral artery (White arrow). The ascending aortic conduit for the vascular access. (Black arrow)

(B) Post-operative CT shows no residual coarctation (Arrow, right) in the stent graft, comparing to pre-operative CT (Arrow, left)

(C) 3-D reconstruction CT shows complete seal of aortic rupture after stent graft placement without endoleak (Arrow), and retrograde filling of left

subclavian artery (Asteric).

References

1. Kron IL, Flanagan TL, Rheuban KS, et al. Incidence and risk of reintervention after coarctation repair. Ann Thorac Surg. 1990 Jun;49(6):920-5; discussion 5-6.

2. Singer MI, Rowen M, Dorsey TJ. Transluminal aortic balloon angioplasty for coarctation of the aorta in the newborn. Am Heart J. 1982 Jan;103(1):131-2.

3. Gravholt CH. Turner syndrome and the heart: cardiovascular complications and treatment strategies. Am J Cardiovasc Drugs. 2002;2(6):401-13.

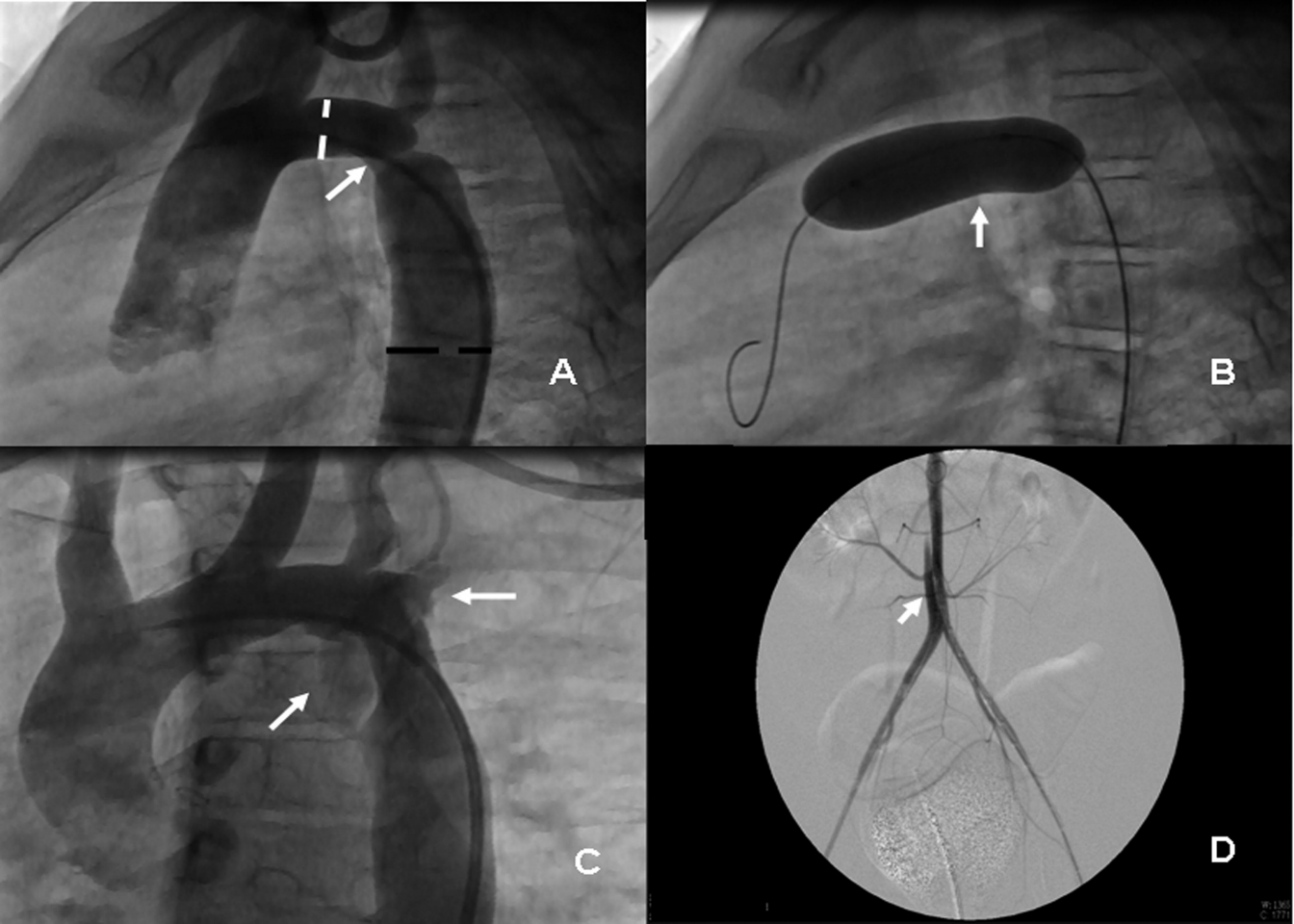
4. Egan M, Holzer RJ. Comparing balloon angioplasty, stenting and surgery in the treatment of aortic coarctation. Expert Rev Cardiovasc Ther. 2009 Nov;7(11):1401-12.

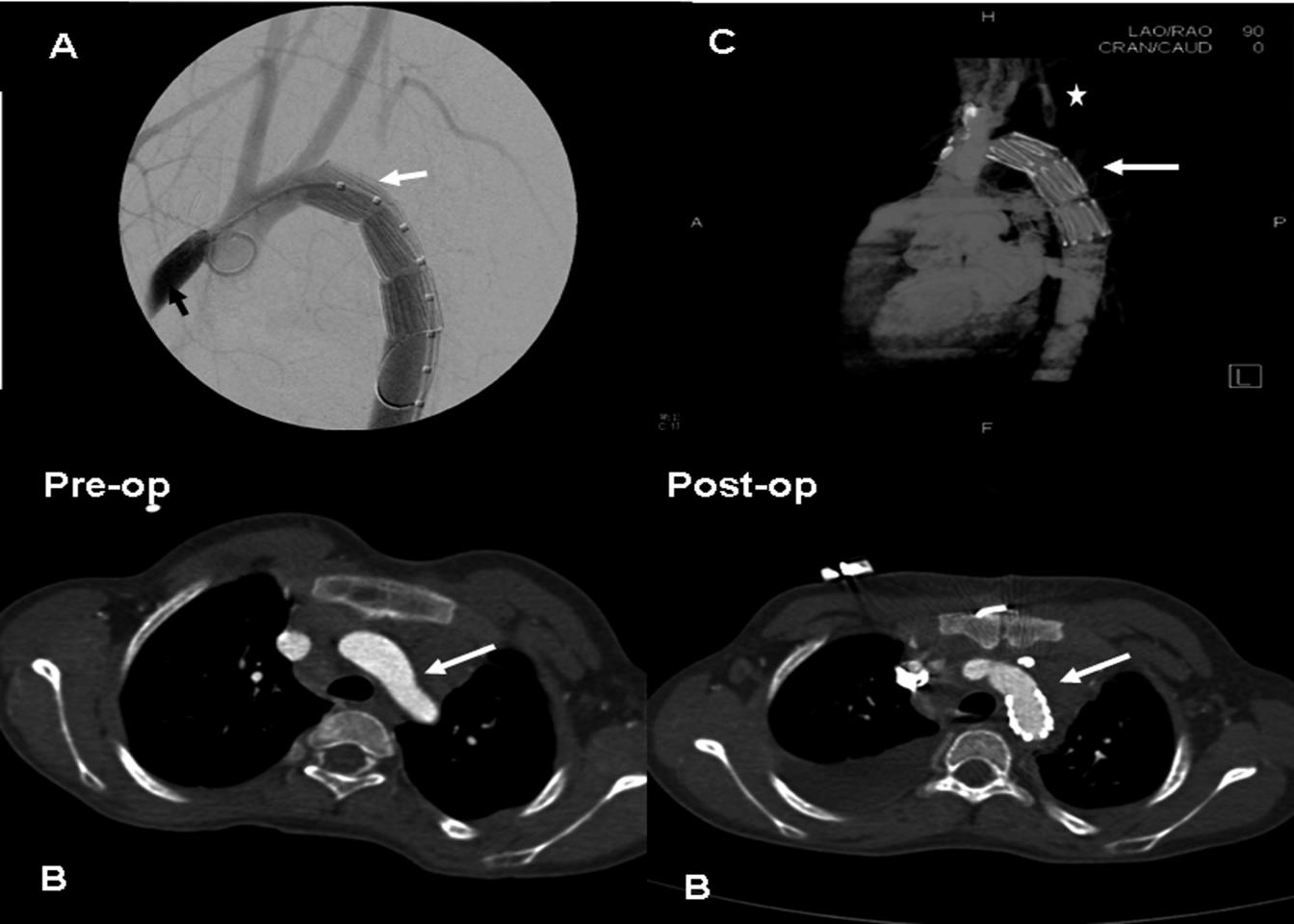
5. Alcibar J, Pena N, Inguanzo R, et al. Stent-graft deployment for aortic rupture after stenting for aortic recoarctation. Tex Heart Inst J. 2007;34(4):453-6.

6. Ozawa A, Predescu D, Chaturvedi R, Lee KJ, Benson LN. Cutting balloon angioplasty for aortic coarctation. J Invasive Cardiol. 2009 Jun;21(6):295-9.

7. Hijazi ZM. Catheter intervention for adult aortic coarctation: be very careful! Catheter Cardiovasc Interv. 2003 Aug;59(4):536-7.

8. Kotelis D, Geisbusch P, Hinz U, et al. Short and midterm results after left subclavian artery coverage during endovascular repair of the thoracic aorta. J Vasc Surg. 2009 Dec;50(6):1285-92.


A
B
C
D


A
C
H
LAO/RAO 90
CRAN/CAUD 0
A
P
F
Pre-op
Post-op
B
B