# Aneurysmal Complications of Coarctation

Is endovascular therapy now the gold standard for repair?

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Coarctation of the aorta is one of the best-known congenital cardiac conditions and describes a narrowing of part of the thoracic aorta, most commonly of the isthmus around the insertion of the arterial duct. This condition accounts for 7% to 10% of all congenital heart lesions,[1] and the presentation varies. Classically, there is severe hypertension from an early age. However, this is not always true, and some patients present at a much later stage with hypertension (or, rarely, with claudication when running long distances).

Over the years, there have been a variety of repair strategies used. Open surgical repair was first described in 1945[2] and is usually performed through a left lateral thoracotomy. There are three standard surgical approaches: resectioning and end-to-end anastomosis, subclavian flap repair, and Dacron patch aortoplasty. An alternative to direct repair that has been developed and used on occasion is ascending aortic-to-descending aortic bypass through a median sternotomy wound. More recently, there has been a move away from surgical approaches for the treatment of these lesions. Balloon angioplasty[3] and stenting of the coarctation site[4] are now standard practice, and in the United Kingdom, they are recognized by the National Institute of Clinical Excellence.

Although coarctation treatment can be successful in the short term, there is a need for careful continued follow-up. The late complications, especially after surgical repair, are well recognized. The most recent study describes the prevalence of restenosis and dilatation at the repair site in 247 patients (largely treated by surgical repair), as assessed by CT and MRI.[5] In this study, restenosis of > 70% at the repair site was present in 31% of patients, with dilatation present in 13% and a discrete aneurysmal segment in 9%. The largest study describing the long-term surgery results of 891 patients (analyzing patients with 1–24 years of follow-up) revealed that 5.4% developed pseudoaneurysms at the site of repair (89.6% of these were in patients undergoing patch aortoplasty, 8.3% after end-to-end anastomosis, and 2.1% after prosthetic graft replacement).[6] Late pseudoaneurysm formation is also a complication of angioplasty and stenting, and its incidence will increase with wider use of these techniques.[7]

These postcoarctation repair aneurysms may occur as a result of widening of the aorta in a fusiform pattern, or they may be more discrete and saccular in nature, most likely secondary to suture line disruption between a prosthetic patch and the aortic tissue. In the absence of any useful measure to predict the risk of rupture in saccular aneurysms, most patients require intervention. In our experience, there is no correlation between size and rupture; therefore, assessment at an early stage should be considered mandatory. For those who have a fusiform aneurysm, we do not know when it should be repaired; however, we tend to agree that this type of aneurysm requires repair, according to standard size criteria in the American Heart Association guidelines.[8] Once repair is considered, there is some controversy on how these aneurysms are best repaired (ie, by endovascular or surgical means). This article describes why endovascular management is the better approach for treating postcoarctation repair aneurysms.

## SURGICAL REPAIR OF POSTCOARCTATION REPAIR ANEURYSMAL DILATATION

Until recently, standard repair strategies involved open surgical revision, with a direct approach to the proximal descending thoracic aorta through a left lateral thoracotomy and replacement of the aneurysmal portion (usually with interposition grafting). Although this approach is thought to provide a durable repair, there are significant challenges. When operating in a scarred area, there is a high risk of bleeding and collateral damage. Significant postoperative morbidity occurs after recurrent laryngeal nerve and phrenic nerve paralysis. The fragile nature of the aneurysm itself means that careful dissection is required before exposing the site for proximal cross-clamping of the aorta; otherwise, rupture occurs, and emergency cardiac bypass is required. Deep hypothermic arrest is often planned for safe access to the arch.

One further complicating factor in surgical treatment is the association with other congenital heart defects. Reports have demonstrated this association in > 90% of patients younger than 6 months of age.[9] The abnormalities are varied, but most commonly include tubular hypoplasia of the aortic arch, ventricular septal defects and patent ductus arteriosus, left ventricular outflow and inflow obstruction, as well as abnormal positioning of the great vessels (primarily transpositions). These associated abnormalities mean that the patients are at significant anesthetic risk. A mortality rate as high as 14% has been noted with surgical repair.[7,10]

Although saccular aneurysms may appear to be a localized area of failure, there may be more widespread disease of the arterial system. It has been postulated that coarctation is part of a more widespread aortopathy.[11] Certainly, there is an association with the bicuspid aortic valve, which would support this view.[9] Given this, there can be no guarantees that further surgical repair will not be needed in these patients. Failure in the late term after surgical repair does occur, and there is often the need for additional repair.

Last, but perhaps most importantly, we must consider the patient's view. Often, the patient has undergone significant thoracic surgery on one or more occasions and is reticent to agree to a reoperation. It is difficult to convince this cohort that another surgical procedure will pay off in the long term.

## AN ENDOVASCULAR APPROACH

Endovascular treatment of thoracic aortic aneurysms has significantly grown in popularity since Dake et al implanted the first thoracic aortic stent graft in 1994.[12] This would seem to be an attractive prospect for treating these patients, as early postoperative complications are rare.

In our unit, we perform procedures under general anesthesia with spinal drain placement for those with an anticipated aortic coverage > 15 cm in length. For patients who require coverage of the left subclavian or left common carotid artery origins, we perform extra-anatomical bypass grafting if a custom-made, scalloped stent graft cannot be used. Most often, the common femoral artery is utilized for access either via a cutdown or, more recently, via a percutaneous approach. The stent is positioned under fluoroscopic guidance and deployed with medication-induced hypotension to avoid "bird-beaking" and stent maldeployment. Once final angiograms are obtained, demonstrating satisfactory stent positioning, the patient is managed in a high-dependency unit environment with a mean arterial pressure of > 85 mm Hg for spinal cord protection. Representative angiograms from such a procedure are shown in Figure 1. CT evaluation is performed at 1 month, with mandatory surveillance for all patients (usually at 2-year periods, when the sac has been excluded and has decreased in size). Pre- and postoperative CT images are shown in Figure 2.

There are concerns regarding the use of endovascular technology in the majority of these patients. These concerns are mainly centered around the effectiveness of stent grafts and whether a long-term seal can be achieved in these patients, many of whom are young and have a near-normal life expectancy.

First, there are significant challenges with these patients when an endovascular approach is considered. The lesion is almost invariably at, or just distal to, the left subclavian artery. This needs to be considered, especially in young patients who may need further surgery later in life. Second, there are commonly significant challenges involved with mismatch of proximal and distal landing zones, as well as tortuosity of the aortic arch and proximal descending thoracic aorta, presumably secondary to remodeling during early life with redistribution of blood flow through the internal mammary arteries to the distal vasculature and as a result of surgery.

Early stent systems were not specifically designed for the challenges of the aortic arch, in terms of their lack of conformability and migration. This presumably explains the small number of patients who underwent endovascular repair in early reports,[13-17] due to a lack of suitability and concern that a long-term seal could not be achieved.

Major advances in stent graft technology have been made more recently. Current-generation devices are smaller and more conformable and can be deployed more safely and reproducibly. Our recent report describes a contemporary study of 13 patients ranging from 27 to 66 years of age.[18] In this study, the initial technical success rate

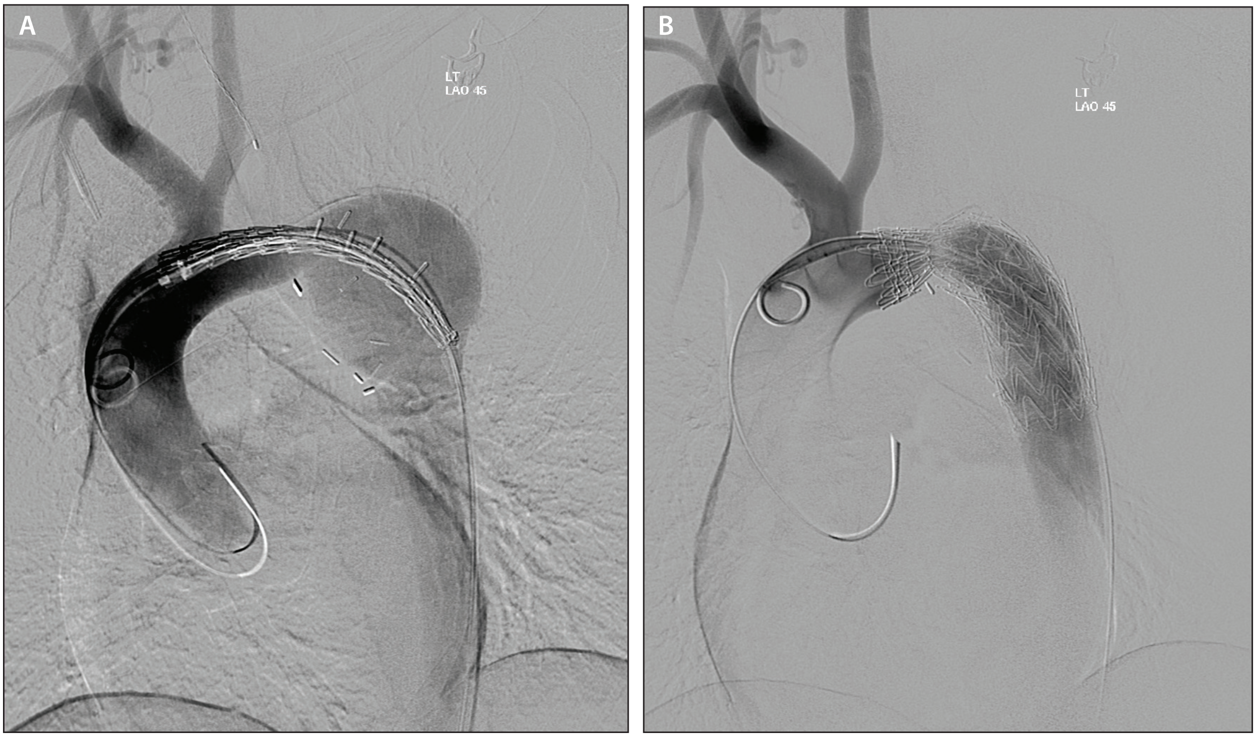


**Figure 1. Intra-arterial digital subtraction angiogram showing a large saccular aneurysm 20 years after Dacron patch coarctation repair. The surgical clips can be seen be seen in place. Note the occlusion of the left subclavian artery from the original surgical procedure (A). Successful exclusion of the aneurysm after placement of a C-TAG stent graft (the residual stenosis that is present was treated with a high-pressure balloon once the covered stent graft was placed) (B).**

was 100% using a variety of stent grafts, with all aneurysms entirely excluded in 10 out of 13 patients. Early CT scans showed type II endoleaks in two patients and a single type Ib endoleak that required distal stent extension.

Despite the significant challenges of location, tortuosity, and device size mismatch, a successful seal can be achieved in the short term. We used a combination of Captivia (Medtronic), Conformable TAG (C-TAG, Gore & Associates), and custom-made stent grafts. This new generation of stent grafts with increased deployment accuracy and fixation, as well as superior conformability, is vital in improving initial technical success. To reduce the risk of paraplegia, it is essential to utilize the shortest aortic coverage possible to gain an effective and durable seal. This can be difficult if multiple stent grafts must be combined to seal a proximal and distal landing zone. Use of the C-TAG device can reduce the need for multiple stents due to a wider range of aortic diameters indicated for use with each stent graft diameter, especially at the lower size range. By utilizing custom-made stent graft systems, the range of aneurysms that can be treated increases. The custom-made Relay device (Bolton Medical, Inc.) has a proximal scallop, which allows for a longer seal zone without the need for extensive extra-anatomical hybrid revascularization of the subclavian or left carotid artery. Again, this device is useful for significant proximal and distal landing zone mismatch, as a taper can be incorporated into the custom-made design. Our experience with this device has grown over the years, and results are promising in the short and medium term.[18]

One significant concern with endovascular stent graft repair in these patients is durability. This is a valid worry, because there have been many reports of late stent graft failure and late mortality rates.[19] However, an examination of large-scale databases has revealed that there is most certainly a differential outcome of thoracic stenting related to initial pathology.[20] It is therefore not a valid argument to compare the results of thoracic stenting for aneurysmal disease with localized pathologies. In the acute situation, some similarities can be drawn from examining the results of aortic transection. These patients have a localized pathology with landing zones that consist of "normal" aortic tissue. In those who survive severe multitrauma, the long-term results of stent placement appear excellent.[21]

In our series, we have reported a median follow-up of 15 months (and now have a median of 2 years experience). Except for the initial distal extension needed for a type Ib

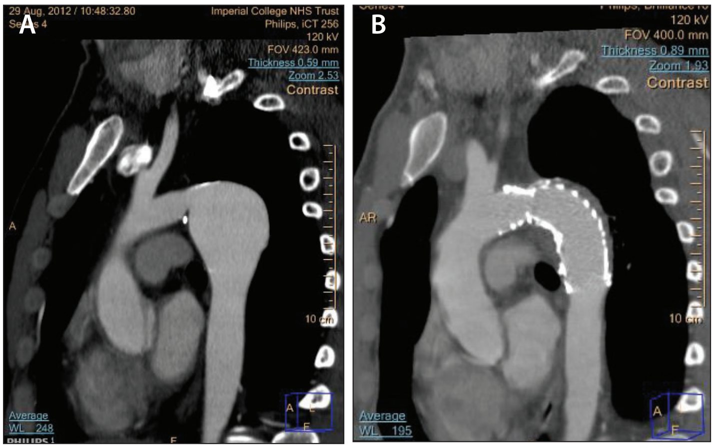


**Figure 2. Preoperative sagittal CT image of the patient in Figure 1 (A). CT imaging of the same patient shows the stent graft in place, with exclusion of the aneurysm and complete regression of the aneurysm sac at 1 year (B).**

endoleak (noted at the time of initial CT evaluation), no patients have required further stent graft intervention, which one may reasonably attribute to careful attention paid to achieving adequate seal and fixation at the landing zones with these newer-generation stent grafts. The majority of the saccular aneurysms have regressed to an impressive degree, so there is no measurable sac.

With further technological advances, we can expect the durability of thoracic stent grafting to further improve. In our unit, we have utilized the thoracic EndoAnchor system (Aptus Endosystems, Inc.) in an attempt to provide a versatile method of providing stable fixation. The long-term outcomes of this system are yet to be reported in the thoracic aorta, but promising results are emerging for the infrarenal segment.[22]

## CONCLUSION

Patients who have undergone interventions for coarctation, particularly with Dacron patch repair, are at significant risk of aneurysmal formation later in life. It is imperative that every patient is followed up and monitored for this complication, ideally using MRI, by adult congenital heart services. Patients who develop aneurysmal dilatation should be considered for repair, as rupture is associated with a high mortality rate. In considering strategies for repair, it should be taken into account that surgical revision carries significant risks, and there is no guarantee of long-term freedom from reintervention. In addition, patients often prefer to explore minimally invasive options. Although endovascular stent grafting was viewed, quite rightly, with caution in the treatment of these young patients, newer stent graft designs offer better conformability, versatility, and durability. They should now be considered as the first choice for treatment of these aneurysms. ■

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1. Padua LM, Garcia LC, Rubira CJ, et al. Stent placement versus surgery for coarctation of the thoracic aorta. Cochrane Database Syst Rev. 2012;5:CD008204.
2. Crafoord C, Nylin G. Congenital coarctation of the aorta and its surgical treatment. J Thorac Cardiovasc Surg. 1945;14:347-361.
3. Lock JE, Bass JL, Amplatz K, et al. Balloon dilation angioplasty of aortic coarctations in infants and children. Circulation. 1983;68:109-116.
4. Suarez de Lezo J, Pan M, Romero M, et al. Balloon-expandable stent repair of severe coarctation of aorta. Am Heart J. 1995;129:1002-1008.
5. Chen SS, Dimopoulos K, Alonso-Gonzalez R, et al. Prevalence and prognostic implication of restenosis or dilatation at the aortic coarctation repair site assessed by cardiovascular MRI in adult patients late after coarctation repair. Int J Cardiol. 2014;173:209-215.
6. Knyshov GV, Sitar LL, Glagola MD, et al. Aortic aneurysms at the site of the repair of coarctation of the aorta: a review of 48 patients. Ann Thorac Surg. 1996;61:935-939.
7. Forbes TJ, Kim DW, Du W, et al. CCISC investigators. Comparison of surgical, stent, and balloon angioplasty treatment of native coarctation of the aorta: an observational study by the CCISC (Congenital Cardiovascular Interventional Study Consortium). J Am Coll Cardiol. 2011;58:2664-2674.
8. Hiratzka LF, Bakris GL, Beckman JA, et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with thoracic aortic disease: executive summary. A report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine. Catheter Cardiovasc Interv. 2010;76:E43-86.
9. Becker AE, Becker MJ, Edwards JE. Anomalies associated with coarctation of aorta: particular reference to infancy. Circulation. 1970;41:1067-1075.
10. Brown ML, Burkhart HM, Connolly HM, et al. Late outcomes of reintervention on the descending aorta after repair of aortic coarctation. Circulation. 2010;122(11 suppl):S81-84.
11. Preventza O, Livesay JJ, Cooley DA, et al. Coarctation-associated aneurysms: a localized disease or diffuse aortopathy. Ann Thorac Surg. 2013;95:1961-1967.
12. Dake MD, Miller DC, Semba CP, et al. Transluminal placement of endovascular stent-grafts for the treatment of descending thoracic aortic aneurysms. N Engl J Med. 1994;331:1729-1734.
13. Ince H, Petzsch M, Rehders T, et al. Percutaneous endovascular repair of aneurysm after previous coarctation surgery. Circulation. 2003;108:2967-2970.
14. Bell RE, Taylor PR, Aukett M, et al. Endoluminal repair of aneurysms associated with coarctation. Ann Thorac Surg. 2003;75:530-533.
15. Preventza O, Wheatley GH 3rd, Williams J, et al. Endovascular approaches for complex forms of recurrent aortic coarctation. J Endovasc Ther. 2006;13:400-405.
16. Marcheix B, Lamarche Y, Perrault P, et al. Endovascular management of pseudo-aneurysms after previous surgical repair of congenital aortic coarctation. Eur J Cardiothorac Surg. 2007;31:1004-1007.
17. Roselli EE, Qureshi A, Idrees J, et al. Open, hybrid, and endovascular treatment for aortic coarctation and postrepair aneurysm in adolescents and adults. Ann Thorac Surg. 2012;94:751-756.
18. Alsafi A, Bicknell CD, Rudarakanchana N, et al. Endovascular treatment of thoracic aortic aneurysms with a short proximal landing zone using scalloped endografts. J Vasc Surg. 2014;60:1499-1506.
19. Bicknell C, Powell JT. Aortic disease: thoracic endovascular aortic repair. Heart. 2015;101:586-591.
20. Patterson B, Holt P, Nienaber C, et al. Aortic pathology determines midterm outcome after endovascular repair of the thoracic aorta: report from the Medtronic Thoracic Endovascular Registry (MOTHER) database. Circulation. 2013;127:24-32.
21. Steuer J, Wanhainen A, Thelin S, et al. Outcome of endovascular treatment of traumatic aortic transection. J Vasc Surg. 2012;56:973-978.
22. de Vries JP, Ouriel K, Mehta M, et al. Analysis of EndoAnchors for endovascular aneurysm repair by indications for use. J Vasc Surg. 2014;60:1460-1467.e1.