

# Contemporary Management of Aortic Arch Aneurysm

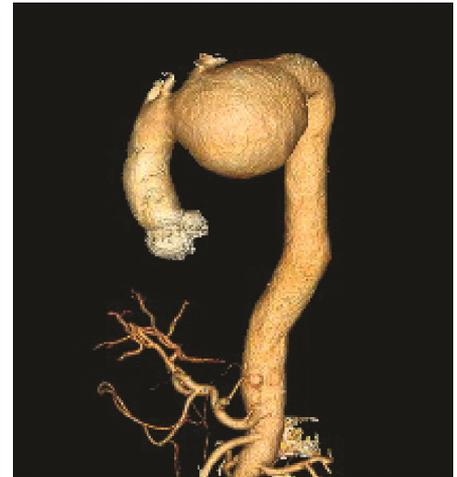


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The management of the aortic arch aneurysm is becoming increasingly complex and multidisciplinary. It has evolved since the first successful repair by DeBakey et al in 1957. After these initial repairs, the improvement in open surgical techniques, cardiopulmonary bypass, anesthesia, and perioperative care were the primary drivers of the decrease in morbidity and mortality associated with repair. The development of endovascular technology has spurred another revolution in the management of aortic arch aneurysms. In this review, we present a current appraisal and description of open surgical, hybrid, and endovascular techniques based on the literature. These techniques are varied and have different advantages and disadvantages, depending on patient anatomy and perioperative surgical risk. We provide an overview of the attributes of each technique and how they may be applied to individual cases. While each technique could not be discussed in detail in this report, it is clear that institutions must be able to proficiently offer the full spectrum of open, hybrid, and endovascular surgical techniques to treat this diverse condition. For low and intermediate risk patients, open surgery remains the gold standard. However, just as improvements in technique, monitoring and perioperative care led to progress in open repair, similar advancements in endograft technology, anatomical customization, and embolic protection will expand the use of endovascular repair. As the management of the condition becomes increasingly nuanced and multidisciplinary, centers must be equipped to offer a variety of techniques with high fidelity and adaptability to each unique patient.

**Semin Thoracic Surg 31:697–702** © 2019 Elsevier Inc. All rights reserved.

**Keywords:** Aortic arch aneurysm, Aneurysmal disease, Endovascular, Hybrid



Aortic arch aneurysm confined to transverse arch with normal diameter DTA.

## Central Message

The management of the aortic arch aneurysm is becoming increasingly complex and multidisciplinary. Treating institutions must be able to proficiently offer the full spectrum of open, hybrid, and endovascular surgical techniques for this diverse condition.

**Abbreviations:** ACP, antegrade cerebral perfusion; CPB, cardiopulmonary bypass; DTA, descending thoracic aorta; EPD, embolic protection devices; ET, elephant trunk; FET, frozen elephant trunk; HCA, hypothermic circulatory arrest; RCP, retrograde cerebral perfusion; SCA, subclavian artery; SCI, spinal cord ischemia; TEVAR, thoracic endovascular stent graft

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Disclosures: The authors have no conflicts of interest and no outside funding was used for this report.

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## INTRODUCTION

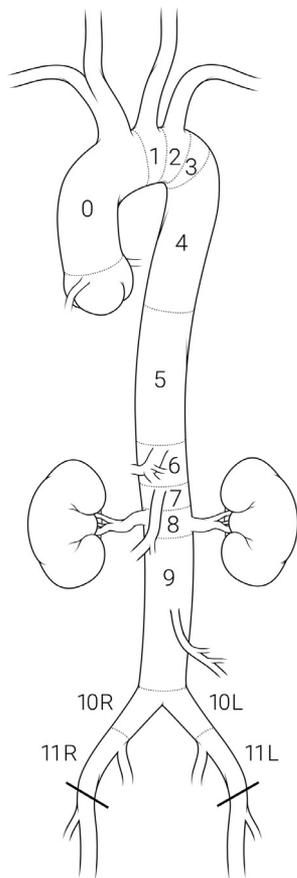
The management of the aortic arch has continued to evolve along with surgical techniques. The initial treatment approach to aneurysmal disease of the aortic arch—like all other aneurysmal disease—had its foundations in open surgical repair. The first successful repair was by DeBakey et al in 1957.<sup>1</sup> After these initial repairs, the improvement in open surgical techniques, cardiopulmonary bypass (CPB), anesthesia, and perioperative care were the primary drivers of the decrease in morbidity and mortality associated with repair. This increased the applicability of open repair to a larger group of patients and encouraged continued innovation to develop techniques to treat even more comorbid patients and difficult anatomy.<sup>2,3</sup>

The development of endovascular technology has spurred another revolution in the management of aortic arch aneurysms. From the first endovascular repair of an abdominal aortic aneurysm,<sup>4</sup> aortic stents grafts have been used in all anatomic

zones of the aorta to aneurysm and other pathology.<sup>5,6</sup> In addition to complete endovascular repairs of the aortic arch, the advent of this technology has allowed for the development of a multitude of hybrid techniques. These hybrid techniques are a combination of open surgical procedures combined with endovascular grafting to achieve the goal of aneurysm repair and/or exclusion. These endovascular and hybrid techniques have made the suitability of repair feasible for an even larger group of patients and aneurysm morphologies.<sup>7</sup> The continued development of new devices is the current and next revolution in management of arch aneurysmal disease.

In this review, we describe the management of aortic arch aneurysm in its current form with available techniques. Contemporary open surgical management, hybrid repair, and entirely endovascular repair will be discussed separately. That being said, the modern management of the aortic arch is starting to blur the lines between these repairs, and individuals managing these patients must be familiar with all options available.

An understanding of the Ishimaru aortic zones is integral in the description and definition of the following techniques. The zones are referenced in [Figure 1](#).<sup>8</sup>



**Figure 1.** The aortic zones of attachment. Adapted from Fillinger et al.<sup>8</sup>

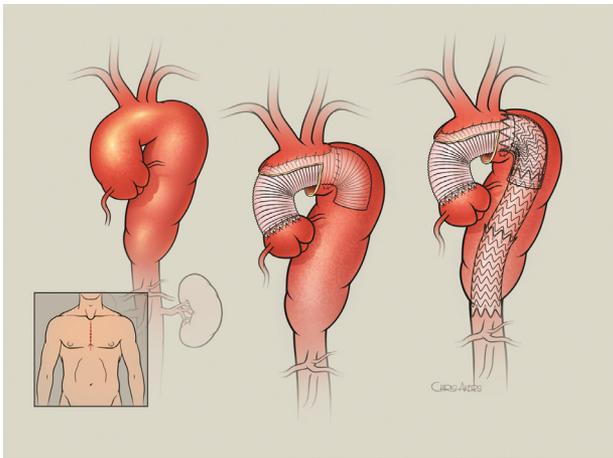
## OPEN REPAIR

### Elephant Trunk

Open total arch replacement with a distal anastomosis in zone 3 of the aortic arch without the use of an elephant trunk (ET) or frozen elephant trunk (FET) is now rarely performed.<sup>9</sup> Its use would be for an aneurysm completely confined to the transverse arch with a relatively normal diameter descending thoracic aorta (DTA) ([Fig. 2](#)). This type of morphology is rare, accounting for less than 10% of transverse arch aneurysms requiring open repair. Total replacement of the aortic arch is now most commonly done with one of the above ET techniques. The supra-aortic branches are anastomosed with either an island patch reconstruction or branched graft. The latter is preferred in younger patients and those with known or suspected connective tissue disorders to mitigate the long-term risk of patch aneurysm. The decision to pursue an ET or FET reconstruction depends primarily on the patient's anatomical presentation and the treating center's experience. The ET is advantageous in that it provides the distal extension required into zone 4 for staged repair of a distal arch aneurysm but does not cover a large length of DTA.<sup>3</sup> The next stage can then be completed with either open or endovascular techniques ([Fig. 3](#)). This technique allows reconstruction of the arch without having to achieve complete exclusion of the distal aneurysm through a sternotomy, which can prove technically demanding in patients with large aneurysms or deep, barreled-shaped chests. Traditionally, the second-stage ET was performed via a modified thoracoabdominal incision with Dacron graft replacement of the diseased segment. Recently, the



**Figure 2.** Aortic arch aneurysm confined to transverse arch with normal diameter DTA.



**Figure 3.** A two-stage elephant trunk procedure. Stage 1 is a total arch replacement and ET. Stage 2 is the endovascular completion with proximal landing zone in ET.

completion second-stage intervention has been accomplished using thoracic endovascular stent graft (TEVAR). In the setting of retrograde deployment, cannulating the ET may offer access challenges, due to crimping of the Dacron ET. Regardless of the length the remaining ET, retrograde stent graft deployment should extend to just distal of the left subclavian reattachment to assure adequate seal.

A FET is done much like an ET, but with deployment of a TEVAR antegrade through the reconstructed transverse arch while still on circulatory arrest and prior to the resumption of CPB.<sup>10</sup> It should be noted that at the time of writing this review, commercially available FET grafts are not yet available in the United States. Series using commercially available stent grafts deployed in an antegrade fashion during circulatory arrest have been reported.<sup>11,12</sup> Advantages are that it can possibly treat distal arch pathology in one stage and—although not the focus of this article—help stabilize true lumen flow in acute dissection to treat malperfusion. It may also reduce the tension on this distal anastomosis and reduce the risk of suture line bleeding, since sealing is now farther distal in the DTA. Disadvantages are its increased length of DTA coverage, with a spinal cord ischemia (SCI) rate of 5%, increased circulatory arrest time, and the need for access to a hybrid operating room.<sup>13</sup> Devices such as Vascutek's Thoraflex Hybrid graft (Inchinnan, Scotland, UK) are available for FET reconstruction. It is specifically made for the technique and features a proximal Dacron graft for open transverse arch reconstruction attached to endovascular stent graft. The endovascular component is deployed distally to proximally in the DTA prior to performing a collared transverse arch anastomosis and reconstruction.<sup>14</sup>

### Cerebral Protection

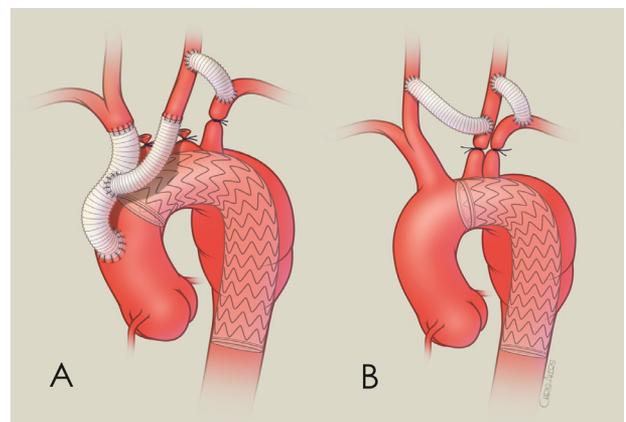
Repair of aortic arch aneurysms was initially performed under deep hypothermic circulatory arrest (HCA) alone. Cerebral perfusion strategies have now evolved over time to decrease the neurologic risks involved with HCA. These

include retrograde cerebral perfusion, and antegrade cerebral perfusion either unilaterally or bilaterally. Head-to-head comparisons have been done in cohort type registries attempting to show superiority of one technique over the other. The results are heterogenous but it is evident that some form of cerebral perfusion is beneficial in comparison to none at all.<sup>15</sup> Our institution employs retrograde cerebral perfusion in cases requiring HCA.<sup>3</sup>

### HYBRID REPAIR

Hybrid repairs have gained popularity as endovascular technology has improved. They offer a potentially less invasive technique for the management of arch aneurysmal disease in patient populations that often have comorbidities precluding direct open repair under HCA. The discussion of hybrid repair of aortic arch aneurysms centers on the strategy of generating a proximal landing zone for an endograft. This landing zone must be in the nonaneurysmal aorta and of adequate dimensions to accommodate the TEVAR and provide endoseal. The landing zone is generated by the debranching of the aortic arch, allowing the stent graft to be placed in an increasingly proximal position. The debranching procedure should be referred to by the last aortic zone that has been excluded.<sup>13</sup> Use of CPB is still required with some cases, but may not be needed depending on the patient's anatomy and the extent of aortic arch debranching needed.

A zone 2 debranching would therefore require bypass or transposition of the left subclavian artery (SCA) via a cervical approach. A zone 1 debranching requires the bypass or transposition of the left common carotid and left SCA via cervical approach. Finally, a zone 0 debranching requires inflow from the proximal ascending aorta via sternotomy with bypasses to all 3 supra-aortic trunks (Fig. 4). A classification of zone 0 debranching procedures have been previously described.<sup>16</sup> This includes a type I hybrid arch repair where the inflow anastomosis for the arch vessel bypasses is sewn directly to the native ascending aorta, and a type II hybrid arch where ascending aorta is replaced and the inflow anastomosis originates



**Figure 4.** Zone 0 hybrid arch repair (type I) (A). Zone 1 hybrid arch repair (B).

**Table 1.** Neurologic Complications, Endoleak and Reintervention Rates of Open, Hybrid and Endovascular Treatment of Aortic Arch Aneurysm

	Stroke	SCI	Endoleak	Reintervention
Elephant trunk	3–5%	1–2%	–	4.3%
FET	3–9%	5–7.9%	–	21% (70% of these planned second stage)
Hybrid repair	7.3–8.9%	3.6–5.5%	15–20.1% (12.9% Type 1)	13% (>90% for type 1 endoleak)
Branched arch grafts	15.8–20% (5.3–7% major stroke)	0–6.0%	14–28.9% (7.0–13.2 type 1)	10.5–12%
Parallel grafts	4.2–17%	7.7%	10.5–23%	11.4% at 5 years

from the ascending aortic graft. The type, diameter, and length of TEVAR used to complete the repair would then depend on the patient's anatomy and institutional preferences (Table 1).

The advantages of hybrid repair are that it does not necessarily require CPB and HCA. Also, other than a zone 0 debranching, it can usually be done through a less-invasive cervical approach. It is constrained, however, by the size limitations of available TEVAR grafts—especially if the ascending aorta and proximal arch are ectatic. The cervical approach is not without potential complications and carries with it a risk of lymph leak, graft infection, and nerve injury.<sup>13</sup> The Achilles' heel of the hybrid repair remains the increased risk of stroke from manipulation of the supra-aortic vessels during debranching and transgression of the aortic arch during TEVAR placement. There is also the risk of retrograde type A dissection (6–11%), SCI, and access site complication during TEVAR.<sup>17</sup>

Extra-anatomic arch reconstruction to facilitate these procedures, however, does demonstrate good long-term bypass patency. In a series of 143 bypasses, 5-year patency was 92%. Operative mortality was 0.7%, nonfatal stroke occurred in 1 patient (0.7%), and TIA in 2 (1.4%). Six (4.3%) late occlusions occurred (all carotid-subclavian bypasses) of which 3 were asymptomatic and 3 resulted in arm ischemia.<sup>18</sup>

We believe another hybrid type repair of the arch deserves mentioning. It is unique in that it combines the advantages of open repair with that of a new endograft technology. Where to place this repair in the spectrum of open-hybrid-endovascular techniques speaks to the fluidity of the field and the importance of understanding all available treatment options available. It has been described as a zone 2 total arch replacement. It employs a distal anastomosis in zone 2 of the transverse arch, and a landing zone within the Dacron graft for a TEVAR proximal to this. The left SCA is then revascularized using a GORE TAG Thoracic Branch Endoprosthesis (TBE), a device that will be discussed in the next section.<sup>19</sup> Alternatively, the left SCA could be revascularized with bypass or transposition after conventional TEVAR. Advantages of this technique include a more proximal distal anastomosis, which makes it easier to control anastomotic bleeding, one less supra-aortic vessel anastomosis to complete while on HCA, a decreased chance of injury to the left recurrent laryngeal nerve, and the option of staging any DTA repair, which may decrease SCI risk.<sup>20</sup>

## ENDOASCULAR REPAIR

### Single and Dual Branch TEVAR

The progression of TEVAR technology and the success of fenestrated and branched endovascular repair of the perivisceral aorta have, ultimately, led to the development of branched and fenestrated aortic arch grafts. The first iteration of this approach was performed in 1999.<sup>21</sup> Many iterations of these devices have been developed by companies with known expertise in the manufacture of aortic endografts. Currently, there are no FDA-approved branched or fenestrated grafts available to patients in the United States. There are devices undergoing investigational device exemption studies, but there are not yet published data. There are 2 devices approved for use in the European market that are available for custom order.<sup>13</sup>

The GORE TBE graft (Flagstaff, AZ) is currently under investigational trial in the United States. It is based on their cTAG thoracic endograft and features a retrograde portal that allows the deployment of a bridging stent to revascularize the left SCA or innominate artery in zone 2 or zone 0, respectively. Initial results from feasibility trials look promising, with no perioperative death or stroke.<sup>19</sup> Medtronic (Minneapolis, MN) also has a feasibility study done for a single-branched aortic arch device called the Mona LSA, based on its Valiant platform. The small trial showed no perioperative death or major stroke.<sup>22</sup> Other single-branch devices exist, including the Endospan Nexus (Herzlia, Israel), which uses a different deployment approach. The main body of the device is deployed from the innominate artery into the DTA with right axillo-femoral access. From there an ascending aortic extension module is brought up from the femoral access through a large side hole fenestration and deployed achieving seal in the ascending aorta.<sup>23</sup>

The 2 approved inner multibranch devices available in the European market are the Zenith branched arch endograft (Cook Medical, Bloomington, IN) and the Relay Branched arch endograft (Bolton Medical, Sunrise, FL). Both use antegrade inner branches that allow deployment and then cannulation from the direct puncture and instrumentation of the target supra-aortic vessels (usually the innominate and left common carotid artery). The sizing and technical details of all these devices are beyond the scope of this review but, for the right anatomy, provide an option for total endovascular reconstruction of the aortic arch.

Much like the hybrid techniques, the concern has been the rate of perioperative stroke and risk of retrograde type A dissection. The experience with both of the multibranch devices has shown a periprocedural stroke rate of 11–16%.<sup>24,25</sup> This is in combination to other complications associated with an endovascular repair, including endoleak, migration, maldeployment, and access site complications. Deployment of these grafts in proper position in the ascending aorta can also be difficult because of the length of the stable platform needed, the rotational forces on the deployment system from vessel tortuosity causing possible misalignment, and the angulation of the arch affecting endograft apposition to the inner curve of the ascending aorta.<sup>26,27</sup> Nonetheless, these devices are valuable tools in the treatment of high-risk surgical patients with aortic arch aneurysm. This is especially true in those who have already had an ascending aortic replacement, where stent deployment into zone 0 carries less risk of causing a retrograde type A dissection.

### Parallel Stent Grafts and In Situ Fenestration

Aortic arch parallel stent grafts use bare or covered stents deployed from supra-aortic vessels, either proximally into the ascending aorta (chimney, antegrade flow) or distally into the DTA (periscope, retrograde flow).<sup>13</sup> A conventional TEVAR is then placed alongside this stent in parallel with the goal of excluding flow from the aneurysm sac and preserving flow into the supra-aortic vessel. This forms a “gutter” between the stents, which may allow for continued antegrade flow into the aneurysm sac. The early type 1a endoleak rate is 11%, with persistence past 30 days of approximately half of these.<sup>28</sup> This technique also requires aggressive oversizing of the TEVAR component and may not be possible in patients with large diameter ascending aortas. Stroke and all the other complications specific to endovascular repair are concerns with parallel grafting.

In situ fenestration uses direct mechanical or laser techniques to perforate the fabric of a deployed TEVAR graft, usually from a retrograde approach in the target supra-aortic vessel. This is then dilated and stented to re-establish flow into that branch. It is an off-label technique and without any robust data. It can damage the fabric and wire frame of the TEVAR device, weakening its integrity and causing type III endoleaks. Like parallel grafting, however, it does potentially have a role as an emergent or bailout technique.<sup>13</sup>

### Embolic Protection Devices

There has been a recent interest in using embolic protection devices to try and reduce the rate of potential embolic events during TEVAR deployment. Feasibility studies have shown that the use of embolic protection devices is safe without much increase in procedure time or complexity. Compared to historical controls, there may be a reduction in the amount of emboli and the volume of affected brain parenchyma on diffusion-weighted MRI, especially in the anterior cerebral circulation.<sup>29</sup> There has also been work done demonstrating that air present

in the TEVAR deployment sheath and mechanism may be a possible cause of cerebral embolic events. Therefore, flushing the graft with carbon dioxide or in an underwater bath prior to insertion may decrease the amount of air released.<sup>30</sup> Whether these interventions decrease, the clinically relevant stroke risk remains to be seen. However, they may offer a method to decrease the rate observed during the endovascular management of the aortic arch.

### CONCLUSIONS

The management of the aortic arch aneurysm continues to progress and change. Refinement of open surgical technique has decreased the perioperative morbidity and mortality rate and has opened the possibility of repair to a larger subset of patients with the disease.<sup>3</sup> In addition, the development of hybrid techniques has allowed the treatment of patients without the need for CPB and HCA.<sup>7</sup> The endovascular revolution in the treatment of aortic pathologies continues to forge on and, as the technology progresses, the applicability of these devices to an increased variety of patient anatomies will be realized.

For low and intermediate risk patients, open surgery remains the gold standard. However, just as improvements in technique, monitoring and perioperative care led to progress in open repair, similar advancements in endograft technology, anatomical customization, and embolic protection will expand the use of endovascular repair. As we have seen with the increased applicability of transcatheter aortic valve replacement,<sup>31</sup> a similar revolution may soon come to hybrid and total endovascular aortic arch repair.

The relationship between volumes and outcome in aortic arch surgery cannot be understated.<sup>13,32</sup> As the management of the condition becomes increasingly nuanced and multidisciplinary, one cannot discount the need for this to occur in centers that can offer a variety of these techniques with high fidelity and adaptability to each unique patient.

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