Aortic arch pathology remains a significant clinical challenge. Despite improvements in operative techniques and perioperative care, open repair of aortic arch aneurysms is still plagued by significant morbidity and mortality. Endovascular approaches have significantly altered our approach to aortic arch disease, with the hopes of diminishing complications and mortality. With endovascular repair, however, there are many obstacles that must be overcome in order to achieve successful exclusion of aneurysms in this location. With growing experience the success rates are improving and the complication rates are declining, and with continued dedication toward device and procedure improvement endovascular arch repair will replace open surgery in many clinical scenarios.

**Keywords:** aortic aneurysm • aortic arch aneurysm • branched endograft • fenestrated endograft • hybrid procedure • TEVAR

Aortic arch pathology is a very complex entity and conventional surgical repair is an invasive procedure necessitating arch replacement techniques with cardiopulmonary bypass and deep hypothermic circulatory arrest. Despite the recent advances in perioperative management, surgical techniques and cardiac anesthesia, mortality ranges between 7 and 17% and the rate of neurologic complications, combining stroke and spinal ischemia, ranges from 4 to 12% even in centers of excellence specializing for aortic disease [1,2]. Factors like advanced age and major cardiopulmonary comorbidities increase the rate of complications.

Endovascular repair has widely replaced open surgical approaches for a wide range of thoracic and abdominal aortic pathology. With the surge of endovascular technology the future role of conventional open technique is being questioned. The development of fenestrated and branched devices has made it possible to treat complex aortic pathologies with decreased mortality and hospital stay. Recently, newer adjunct techniques such as parallel grafting, *in situ* fenestration and branched endografting has made feasible the total endovascular reconstruction of the aortic arch, avoiding the need of a median sternotomy or thoracotomy.

The aim of this article is to review the endovascular options for the treatment of aortic arch aneurysms, describe briefly the current techniques and the challenges that are unique for the aortic arch; and evaluate the short- and mid-term outcomes of those repairs.

**Background description of hybrid procedures**

Successful stent-graft deployment and exclusion of aortic aneurysms requires an adequate region of fixation and sealing proximal to the diseased aorta. The length and diameter of this region may vary based on the Indications for Use for specific manufacturers stent grafts, but in general require a parallel segment of aorta that is at least 20 mm in length and free of angulation and thrombus deposition. One critical need when addressing the aortic arch is to maintain perfusion over the arch vessels by either incorporating them into
the repair, or redirecting blood flow to them through an alternate route. Historically, and still in many situations, this is achieved through a combination of open surgical debranching of the great vessels followed by endovascular exclusion of the aneurysm in either a single- or two-staged approach.

The extent of the debranching depends on the proximal extension of the aneurysm according to the Ishimaru classification. The anatomical endograft landing zone map was advocated at the First International Summit of Thoracic Aortic Endografting in Tokyo in 2001 [3]. According to the classification, zone 0 is the ascending aorta from the aortic valve, including the innominate artery, zone 1 from just beyond the innominate artery, including the left common carotid artery (CCA), zone 2 from just beyond the left CCA, including the left subclavian artery (LSA), and zone 3 and 4 beyond the origin of the LSA to the beginning of the descending thoracic aorta.

Extension of the aneurysm into zone 0 makes tentative a revascularization approach in order to maintain perfusion into the supra-aortic vessels. Many techniques have been described to accomplish this, but the most common requires a median sternotomy and the placement of a bifurcated graft in the anterior aspect of the ascending aorta anastomosed to the innominate artery and left CCA [4]. An additional left CCA/LSA bypass or transposition can be made. Bavaria et al. described a similar hybrid repair with placement of a trifurcated graft in the ascending aorta in patients with significant comorbidities, and the early results were acceptable and even encouraging [5]. Endovascular approaches into the zone 0 location are the future of endovascular therapy and represent where significant advances will be made in the ensuing years.

Extension of the stent graft into zone 1 makes mandatory the revascularization of the left CCA which is most commonly done with a right-to-left CCA bypass [6]. This procedure avoids the need and the morbidity of a median sternotomy. Patency rates were reported approximately 88% at 3 years and 84% at 5 years with primary-assisted patency exceeding 90% at 5 years [7]. Depending on anatomic and physiologic variables the LSA may be revascularized or covered by the stent graft.

A significant number of aortic pathologies involve the distal arch adjacent to the LSA. Extension of the stent graft to zone 2 requires partial or complete coverage of the LSA with or without carotid-subclavian bypass or transposition. Patients with hypoplastic right vertebral artery, noncomplete circle of Willis, or LIMA to LAD bypass need LSA revascularization. In patients that require an elective TEVAR with coverage of the LSA, the current Society of Vascular Surgery (SVS) guidelines recommend preoperative LSA revascularization, despite the very low quality evidence [8]. Patients with occluded hypogastric arteries, previous descending thoracic or abdominal aorta coverage need to be considered candidates for LSA revascularization to avoid spinal cord ischemia.

Elephant trunk technique & endovascular elephant trunk completion

The elephant trunk technique was described by Hans Borst et al. [9] in 1983 and is a staged procedure for the reconstruction of the aortic arch and the descending aorta. The ascending aorta and arch are replaced during the first stage with anastomosis of the graft beyond the LSA. The elephant trunk of the graft is hanging in the descending aorta. During the second stage, a left thoracotomy is performed and the proximal anastomosis of the second graft is made with the elephant trunk. The mortality of the second-stage procedure is estimated up to 9.6% in a recent study [10]. In order to avoid the morbidity of an additional open procedure, a second-stage transfemoral endovascular stent-graft placement has been described. The operative mortality of this hybrid approach is reported from 0 to 8.3% and no permanent cases of paraplegia were reported [11].

Frozen elephant trunk technique

The frozen elephant trunk (FET) technique is a modification of the conventional elephant trunk technique described by Borst and colleagues in 1983. This is a single-stage procedure, requiring a median sternotomy with cardiopulmonary bypass where the ascending aorta and arch being replaced and a stent graft is being deployed through an antegrade fashion into the descending aorta through the open aortic arch. Results of the FET are being published with a mortality rate 3–12%, stroke rate 0–16% and spinal cord ischemia 0–24% [12–14].

In a recent meta-analysis Koullias et al. proposed a new hybrid classification system. In hybrid type I repair, which incorporates the endovascular elephant trunk and FET technique, the arch is replaced through an open approach and the endograft extends the treatment area by exclusion. The role of the endovascular component is secondary. In hybrid type II repair, a surgical graft is used for debranching and an endoluminal stent graft is used for exclusion. The role of the endovascular component on this approach is primary. A total of 15 studies with 463 patients were included in this meta-analysis. The overall mortality was 8.3%, the stroke rate was 4.4% and paraplegia rate was 3.9% [15]. The development of hybrid stent grafts in Europe and Australia, like the E-vita Open Plus® (Jotec UK Ltd, Evesham, UK) and Thoraflex® Hybrid graft (Vas-
ctec; Vascutek Ltd a Terumo Company, Renfrewshire, Scotland) have helped to simplify and standardize the FET procedure. Both devices have similar characteristics. The proximal component is made by woven polyester covered with gelatin, the arch branches are already incorporated in the case of Thoraflex, which has a proximal tetrafurcated component, and the distal part is a stent graft which is already presewn in the proximal graft. Potential advantage of those hybrid stent-grafts is the completion of the procedure in a single stage with decreased time of extracorporeal circulation. Results so far are coming from small case series and case reports, and the experience in the USA is limited [16,17].

Aortic arch challenges for a total endovascular repair
Greenberg et al. [18] highlighted some of the early complications associated with endovascular therapy in the distal aortic arch. While deploying a first-generation device across the LSA in one patient, the proximal bare stent of the endograft damaged the greater curvature of the distal arch due to the lack of conformity of the endograft to the curvature of the arch; the endograft ‘stood upright’ in the angulated portion of the aortic arch, which is now termed ‘bird beaking’. It became obvious very early that in order to develop a device and the techniques to accurately and safely deploy and endograft in the aortic arch it is very important to understand the anatomic peculiarities of that area.

The diameter of the ascending aorta is larger than that of the abdominal aorta and the inherent 270-degree angulation between the arch and the descending aorta makes it an extremely difficult target. On the way up to the aorta, the stent graft floats like a windsock under the influence of the projectile flow from the heart [19] and the endograft migration during deployment can lead to significant type I endoleak or collapse of the device with devastating complications. There are some methods of minimizing this effect, including adenosine-induced transient cardiac arrest [20], pharmacologically induced hypotension or ventricular fibrillation [21], transient hypotension by inflow occlusion with intra-atrial balloon or vena cava occlusion [22,23]. More recent endograft designs can ameliorate some of these issues allowing for more controlled deployment without the need for pharmacologic pressure manipulation during graft deployment, at least in the more distal arch.

The curvature and the proximity of the arch vessel ostia make it even more difficult to obtain an adequate proximal sealing zone. A precise deployment is mandatory in order to establish ante grade flow to the branches of the arch through the fenestrations/branches. The proximal fixation needs to be immediately distal to the sinotubular junction otherwise the coronary vasculature will be occluded. Many times the sheath needs to be advanced through a diseased aortic valve into the left ventricle. An additional aortic arch challenge that should be mentioned is the presence of a mechanical aortic valve. The presence of a mechanical valve can be prohibitive for placement of a graft in which crossing of the valve with the delivery system is mandatory. Future device development will include systems that can be placed just distal to the sinotubular junction without requiring sheath and wire crossing of the aortic valve. These improvements are already underway. In addition, additional aortic arch challenges included the potential risk of retrograde dissection in patients with connective tissue disorders, intramural hematoma and short proximal landing zones. Device characteristics of proximal bare stents and barbs and technical aspects like balloon angioplasty increase the risk of retrograde dissection, which is a feared complication after TEVAR and occurs in 1–3% of patients [24].

Device delivery is also challenging since adequate access is required in order to accommodate the diameter of those devices. Adjunct procedures like iliac conduits are not uncommon to occur. An alternative approach of TEVAR procedure is being described through transapical access. The fast growing experience in transapical valve implantation (TAVI) has led to the implantation of this technique as an alternative to direct access by surgeons familiar with this approach in patients with difficult access anatomy or previous sternotomy. Potentially more clear indications, along with assessments of risks and benefits, will be developed in the coming years [25]. The first-generation devices had decreased trackability along the aortic curvature, and several attempts were made in order to correct the last one like widening the interval between the Z-stent skeleton or adding some internal angulations to the devices and delivery sheaths [26]. Finally, the loss of torque control of the device during delivery inside the tortuous vessels and the need for catheter wire manipulation within an atherosclerotic area of the aorta is a predisposing factor for a stroke [27].

Experience with fenestrated stent grafts in the treatment of arch aneurysms
Progress has not come without obstacles, so placing endografts more proximal in the aorta presents complex challenges as mentioned above. Creative solutions have emerged to address those challenges and one of them is the application of fenestrated endografts, home made devices or in situ graft fenestrations. A wide variety of arch pathology has now been reported, including aortic dissections, aneurysms, penetrating ulcers, tran-
sections and pseudoaneurysms, but our focus is going to be in the aortic arch aneurysms (Table 1).

In April 2008 a clinical study of another TEVAR device, the Najuta® endograft (Kawasumi Laboratories, Tokyo, Japan) began in Japan. The Najuta is a fenestrated device that preserves blood flow through branches of the aortic arch and is suitable for distal arch aneurysms. Kawaguchi et al. in 2008 reported their experience with fenestrated endografts in the treatment of distal aortic arch aneurysms. Between 1995 and 2008, nearly 1100 endovascular procedures were performed to treat thoracic aneurysms. Of these, 682 were performed in Tokyo Medical University, including 474 cases without dissection. Approximately 288 patients were treated with fenestrated grafts. The initial success rate was 95.2% (defined as absence of type I and III endoleak). Complications occurred in 26 patients (3.8%) and were the result of cerebral infarction caused by embolism. Among the 26 patients who had a stroke, a fenestrated graft was used in 16; this provides a stroke rate in patients who required a fenestrated device in the aortic arch of 5.5%. There were five serious strokes (1.7%) and the rate of paraplegia due to spinal cord ischemia was 2.6%. Results were improved over time, which represented growing surgeon experience with no incidence of infarction since 2007 [28].

Azuma et al. [29] from Tokyo Women’s Medical University, Tokyo, Japan, evaluated in a 2-year clinical trial 393 patients who underwent endovascular repair with the next-generation Najuta graft. Of the patients, 340 had degenerative aneurysms. In a previous clinical trial the criteria for patient inclusion was a proximal landing zone of >10 mm. Aortic diameter needed to be <42 mm since the largest available endograft was 45 mm. A precurved, fenestrated device was used, by suturing vascular graft material (expanded PTFE) to a self-expanding Z-stent. All grafts were custom made for each patient, and according to the author 19 types of curved stent skeletons and 8 types of graft fenestrations were available. The second-generation device had larger Z-stent and improved curvature supporting struts to accommodate better in the curvature of the aortic arch and avoid migration. A ‘through and through wire’ technique was used as guidance. This method involves a right brachial access, insertion of guide wire into the innominate and capturing the wire with a snare catheter delivered through femoral access. During deployment, pushing forward against the greater curvature prevents migration and displacement of the device towards the blood flow [30].

The precurved shape of the stent graft automatically controlled the rotation of the device and technical success was reported 99.2%. The procedure was not successful in three patients due to poor access. In all cases the proximal landing zone was between zones 0 and 2. The hospital mortality rate was 1.5%, cerebrovascular accidents occurred in seven patients (1.7%) and permanent paralysis in three (0.76%). Retrograde aortic dissection occurred in three patients, all of them had previous aortic dissection and one patient had a collapse of his graft 4 days after the procedure due to small and very curved aortic arch. The LSA was intentionally covered in 281 patients without revascularization and in 17 with concomitant revascularization. The authors found that with the second-generation endograft there was a significant reduction in cerebrovascular accidents and endoleaks despite the shorter landing zone and the more proximal extension towards the aortic valve.

In another study, Murphy et al. [31] described the technique of fenestrated endografting for the treatment of aortic arch aneurysms. A total of 131 patients were treated with TEVAR for thoracic aortic disease – 57 patients had pathology extension in zones 0–2 and 12 patients in zones 0 and 1. Graft fenestrations were used for the LSA in five patients with in situ laser technique. In order to reestablish flow to the covered vessel, a 2.3 mm Turbo Elite® laser ablation catheter (Spectranetics, CO, USA) was used against the endograft fabric. A 7-Fr sheath was used for support of the catheter against the graft and for protection of the vessel. After the creation of the fenestration the opening was balloon-

<table>
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<th>Author (year)</th>
<th>No. of patients</th>
<th>Mortality</th>
<th>Neurological complications</th>
<th>Ref.</th>
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<td>Kawaguchi (2008)</td>
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<td>62% 5-year survival</td>
<td>CVA (%)</td>
<td>5.5</td>
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<td>Murphy (2012)</td>
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<td>3.4% in-hospital mortality</td>
<td>SCI (%)</td>
<td>10.3</td>
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<tr>
<td>Azuma (2013)</td>
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<td>1.5% in-hospital mortality</td>
<td></td>
<td>1.7</td>
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<tr>
<td>O’Callaghan (2014)</td>
<td>33</td>
<td>7% in-hospital mortality</td>
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<td>7</td>
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CVA: Cerebrovascular accident; SCI: Spinal cord ischemia.
profiled and stented with flaring for reinforcement and adequate sealing. A homemade-branched Talent® graft was used for the innominate artery fenestration in one patient. An 8-mm ePTFE graft was sutured to laser created fenestration ex vivo. The branched endograft was precannulated with a stiff glidewire and reloaded to the sheath for delivery into the arch. One more patient had a left CCA stenting with homemade fenestrated graft in a similar fashion. Neurological complications were the most common adverse event in total eight patients (13.8%) and spinal cord ischemia in two patients both of them with LSA coverage (3.4%). Five of the six strokes were embolic in origin. There was no association between stroke and branch level involvement or type of revascularization (open debranching vs endovascular revascularization). The presence of aortic plaque on preoperative CTA was a negative predictive factor and that was present in two out of six strokes.

In order to evaluate our experience and the midterm results with fenestrated, chimney, scallop configurations in Cleveland Clinic [32], we reviewed all TEVARs between January 2004 and February 2013, that endovascular sealing was extended within the aortic arch along with concomitant stenting of a supra-aortic vessel. Of 767 TEVARs, 33 patients (4%) met the inclusion criteria. 18/33 patients had a noncustom repair (chimney or scallop) and 15/33 had a custom (fenestrated device) repair. Overall 39% of the chimney/scallop groups were emergencies and obviously all 15 custom fenestrated repairs were in a nonurgent setting. In the fenestrated group there were four respiratory complications, one renal complication, one surgical site infection, one conversion to open repair and one death from middle cerebral artery occlusion. In the chimney group there were three deaths, one of them occurred due to loss of brain function after bilateral cerebellar stroke. There were four branch-related problems, one in the fenestrated group and three in the chimney group. The single failure in the custom group was intentional exclusion of the left subclavian artery due to type Ia endoleak. Overall there was a trend toward superior patency overtime in the fenestrated group but without statistical significance. We found that even there was a trend for improved durability in patients with fenestrated repair; both groups were at high risk for perioperative and short-term neurological complications and retrograde dissection. This is why we abandoned the fenestrated technique in this territory and we investigated other solutions, like branched endografts.

**Branched stent graft for endovascular repair of aortic arch aneurysm**

The fundamental concepts for the development of a branched device in order to treat aneurysms of the aortic arch is to create a device which is stable to the respiratory, pulsatile and hemodynamic variations and also accomplish proximal and distal sealing while maintaining intraoperative and postoperative perfusion to the aortic branches.

The first branched device for the aortic arch was created by Inoue in 1996 [33]. The graft was placed in a 51-year-old female patient with type B dissection with a large entry tear just beyond the left subclavian artery. The straight Dacron graft with the side branch was successfully and uneventfully implanted. This initial experience was followed by several other attempts by Inoue and colleagues as an attractive alternative to open repair. The device was consisted by a unibody graft with side branches that were captured with a snare and pulled into the aortic trunk vessels. Primary success was only 60% with major cerebrovascular complications [34].

An innovative technique with a modular branched stent graft for endovascular repair of aortic arch aneurysms described in 2003 by Chuter et al. The prosthesis was consisted by two components: a bifurcated proximal component and a tubular distal component. Both were made by polyester material and were supported by stainless steel stents in Z-configuration. A carotid–carotid bypass was performed with reimplantation of the left subclavian into the left CCA. The proximal bifurcated component was delivered antegrade through the right CCA and the short aortic limb was cannulated through femoral access. The second sheath was introduced followed by deployment of the tubular distal component and the procedure was completed with ligation of the left CCA. This method has fallen out of favor due to delivery issues through the right CCA/innominate artery, and the high stroke and mortality risk [35].

Haulon et al. [36] recently presented a multicenter evaluation for the endovascular exclusion of arch aneurysms with branched endografts. Ten high-volume centers across the world participated in this retrospective study. Patients were treated with a branched endograft made by Cook Medical® (Bloomington, Ind., IN, USA) designed to each patient’s anatomy (Figure 1). Anatomic criteria included arch aneurysms and chronic dissections, no prior aortic valve replacement (biological or mechanical valves), length of ascending aorta ≥50 mm (measured from sinotubular junction to origin of innominate artery), sealing zone within the ascending aorta ≥40 mm length and 38 mm diameter, innominate artery 20 mm in diameter and ≥20 mm in sealing zone length, and iliac access able to accommodate 22F or 24F sheaths.

Each graft had 2 internal side branches (Figure 2) with an enlarged internal opening at their distal ends. The distal parts of the endograft were wide and flexi-
Figure 1. (A) Computed tomography scan of a patient with an aortic arch aneurysm (arrow). The patient was considered high risk for conventional surgery and was considered for an endograft approach. (B) The patient underwent endovascular repair of the aortic arch aneurysm with a branched aortic endograft. The graft had two branches to incorporate the inominate artery (arrow) and the left carotid artery; and the patient underwent a carotid–subclavian artery bypass preoperatively to preserve flow to the left subclavian artery as the stent graft occluded its origin.

Figure 2. Representative images of the arch branched endograft constructed by Cook Medical. (A) The graft is designed with two branches to allow for incorporation of the inominate and left carotid (or left subclavian) arteries. After deployment of the device there is continued flow to the arch vessels through the system which allows time for cannulation of the branches and further stenting. (B) The branches are placed internally and allow for mating with additional stent grafts that are extended to the target vessels.
Endovascular treatment of the aortic arch  Perspective

ible, but the middle part (the side branch part) was narrow and straight preserving like that perigraft flow and facilitating the cannulation. A curved introducer was used for the delivery, and the innominate artery component was a custom made low-profile graft. Covered stents were used for the component of the left CCA. Briefly, 38 patients were treated with a double inner branch aortic arch endograft. Technical success was 84.2% with secondary procedures performed in 10.5%. The rate of cerebrovascular complications was 15.8% and no aneurysm related death was detected during the 12-month follow-up period. A significant learning experience was appreciated.

Conclusions
The concept of the endovascular repair of aneurysms of the aortic arch is being influenced by the application of this technology in the area of the abdominal aorta with the usage of fenestrated/branched devices. We have to realize although that the arch is a complete different area form anatomic and physiologic standpoint compared with the juxta or pararenal aorta. Conventional surgical repair remains the gold standard repair for the aortic arch despite the substantial rate of morbidity and mortality. More data and clinical experience with those techniques need to be collected and for now the arch branch procedure should be considered only in patients deemed very high risk for conventional open approach and only in high-volume specialized centers.

Future perspective
The experience with complete endovascular repair of the aortic arch remains in its early stages. Significant improvements to this devastating disease process have occurred since the development of endovascular therapy. As with endovascular approaches to other complex aortic regions, such as the visceral aorta, we have observed the evolution from open repair to hybrid procedures, to total endovascular repair. We are observing this same trend with the approach to aortic arch disease. Hybrid approaches to aortic arch replacement are growing routine. Early outcomes for devices specifically designed to tackle arch pathology are undergoing investigation. Over the next 5 years we will see an increase in the number of devices evaluated in this arena. We will continue to hone the procedural approach to limit the perioperative morbidity, in particular stroke. Durability assessment will begin to unfold, and great efforts will be placed on developing ‘off-the-shelf’ designs that will allow for ready access for patients in urgent or emergent situations. Beyond 5 years, endovascular approaches to the arch will likely replace the majority of open surgical repairs performed.

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M Eagleton is a consultant for Cook Medical (Bloomington, IN, USA) and Bolton Medical (Sunnyside, FL, USA). The authors have no other relevant affiliations or financial involvement with any organization or entity with a financial interest in or financial conflict with the subject matter or materials discussed in the manuscript apart from those disclosed. No writing assistance was utilized in the production of this manuscript.

Executive summary

Background & hybrid approaches
• Arch pathology remains a devastating disease, and open repair carries with it a significant morbidity and mortality.
• Endovascular repair of aortic arch disease has evolved significantly over the past few years.
• Currently, the majority of approaches utilize a hybrid approach of surgical debranching of the great vessels followed by endovascular exclusion of arch aneurysms.

Fenestrated/branched endografting for the aortic arch
• Complete endovascular approach to arch disease is in its early stages. Early clinical assessment demonstrates that endovascular exclusion of arch aneurysms with continued perfusion of the great vessels is technically feasible. Attempts at maintaining arch branch perfusion may be performed through a variety of techniques including the use of fenestrations (custom-designed and those created in situ) and branches.
• The major risks of repair include an incomplete exclusion of the aneurysm due to an inadequate seal, and the perioperative development of a stroke. These risks significantly decline with surgeon experience and procedural and device improvements.
References

Papers of special note have been highlighted as:
• of interest; •• of considerable interest


•• Represent a key manuscript describing the use of the elephant trunk procedure.


•• Represent a key manuscript discussing open surgical repair of the ascending and aortic arch.


• Outlines a hybrid arch repair classification system.


• Represents a large series of patients enrolled in a clinical trial with an endograft device to treat aortic arch aneurysms.


• First large, global series evaluating the initial outcomes of total aortic arch replacement with aortic endografts.