

GENERAL PRINCIPLES

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Historical perspective – the evolution of aortic arch surgery

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Introduction

The challenges involved in aortic arch repair are such that the field of aortic arch surgery has existed for scarcely more than 60 years. However, the history of its foundations – the development of our understanding of aneurysms and aortic anatomy, and the rise of techniques and technology for cardiovascular surgery – can be measured in millennia.

Aneurysms from the ancient world to the nineteenth century: diagnosis and non-surgical treatment

It is clear that the ancient Egyptians suffered from aortic disease; signs of aortic atherosclerosis have been found in Egyptian mummies [1]. There is also evidence that the ancient Egyptians were aware of the existence of aneurysms, at least those of the peripheral type. The Ebers Papyrus (Figure 1.1), which was written in 1500 BC or earlier and is probably the most well-known ancient Egyptian medical document, appears to describe an aneurysm as '... a swelling of vessels ... it is hemispherical and grows under thy fingers at every going [i.e. it pulsates], but if separated from his body it cannot become big and not come out [i.e. diminish] ... it is a swelling of a vessel ... and it arises from injury to a vessel' [2]. However, ancient Egyptian physicians could do little to treat aneurysms or many other serious ailments, and their frequent frustration in the face of these conditions is revealed in another passage from the Ebers Papyrus: 'A suffering person is not to be left without help: go in to him, and do not abandon him' [3].

Ancient Asian civilizations may also have been aware of aneurysms. For example, in India, between 800 and 600 BC, Indian surgeon Sushruta described peripheral aneurysms in his work, *Samhita*, as localized, pulsatile swellings in blood vessels. Sushruta recommended treating these swellings with compression, cauterization, or excision [4].

In the second century AD, the Greek physician Galen wrote what some believe to be the first true description of an aneurysm: 'When the arteries are enlarged, the disease is called an aneurysm. ... If the aneurysm is injured, the blood gushes forth, and it is difficult to staunch it' [5]. (Because exact translations are not always available for medical terms in ancient languages, there is some debate as to whether the pre-Galenic texts discussed here really describe aneurysms and not some other disease. For example, the word translated as 'vessels' in the Ebers Papyrus is metu, which was used to refer not only to blood vessels but also to muscles, nerves, or any other long, thin body structure [4].) Also in the second century, the Greek surgeon Antyllus produced the first known writings on the causes of aneurysms. He distinguished between aneurysms caused by trauma and fusiform or cylindrical



Figure 1.1 A passage from the Ebers Papyrus, which may contain the first known record of aneurysmal disease. The manuscript appears to state that aneurysms should not be treated surgically, but only by incantation. It also repeatedly admonishes the physician not to abandon the patient.

aneurysms, as they are called today, caused by syphilis or other chronic diseases. Antyllus described treating these aneurysms with proximal and distal ligation and evacuation of the sac – a technique that remained the standard of care until the eighteenth century [6].

Aortic aneurysms do not appear to have been identified as such until the Renaissance era, when the dissection of corpses began to become an acceptable practice, at least in some circles. In 1542, prominent French physician Jean-Francois Fernel (who has been credited with, among other things, coining the terms 'physiology' and 'pathology') published his work De Externis Corporis Affectibus, in which he distinguished between 'external' aneurysms (i.e. aneurysms of the peripheral vasculature) and 'internal' ones (i.e. aneurysms of vessels within the chest and abdomen, including aortic arch aneurysms). Fernel's contemporary, University of Montpellier chancellor Antoine Saporta, described the pulsatility of aortic aneurysms, thus distinguishing them from tumors, and he also described the symptoms of fatal aortic rupture. Illustrations of aortic arch aneurysms in particular appeared in several books written in the sixteenth century and thereafter [4].

Between the sixteenth and nineteenth centuries, many theories were proposed about the genesis of aortic aneurysms, and some of these theories were later substantiated. For example, several prominent physicians and scientists suggested that syphilis played a causal role in many aortic aneurysms; in the seventeenth century, two Italians, anatomist Giovanni Lancisi and surgeon Marcus Aurelius Severinus, both described the weakening of vessel walls in syphilitic persons. This theory was substantiated in 1876, when Francis Welch published his series of post-mortem examinations of patients with or without aortic aneurysms. Of 53 patients with aneurysms, two-thirds had clear signs of syphilis, whereas all but one of the 106 non-syphilitic patients he examined had no aortic dilatation [7]. Although there was considerable resistance even to the discussion of this theory at the time, the notion that tertiary syphilis caused aortitis that led to the formation of aortic aneurysms eventually became commonly accepted. The discovery of penicillin in 1928 made syphilitic aneurysms a rarity.

Other useful theories about the origin of aneurysms were also introduced before the twentieth century. Fernel, in the 1600s, correctly theorized that fusiform or cylindrical aneurysms caused by degenerative disease resulted from the simultaneous dilatation of all layers of the artery, rather than the dilatation of individual layers as some of his contemporaries asserted. Also, Lancisi posited traumatic and congenital origins for some aneurysms. In the nineteenth century, another Italian anatomist, Antonio Scarpa, suggested atherosclerotic degeneration of vessels as a cause of some aortic aneurysms [4].

Awareness of aortic arch coarctation also arose in the eighteenth and nineteenth centuries. This problem was

first described by Johann Friederich Meckel in 1750 and by Morgagni in 1760 (although Morgagni described it as a localized constriction of the descending aorta) [8,9]. However, it would be almost 200 years before any attempt at surgical intervention was made.

The advent of surgical treatment for aortic arch disease

For centuries, total bed rest, starvation, and dehydration were the standard treatment for aneurysms. External aneurysms were sometimes treated by direct compression with bandages, cauterization with hot irons, limb amputation, and ligature of parent arteries. Internal aneurysms, however, remained untreatable; sixteenth-century surgeon Ambrose Paré wrote that 'the aneurysms which happen in the internal parts are incurable' [10].

In the late eighteenth century, some physicians began to advocate treating aortic aneurysms by introducing heated needles into the sac to stimulate thrombosis. However, the results were unpredictable, and the technique fell out of favor for some time [11]. Then, in 1864, Charles Hewitt Moore of London's Middlesex Hospital introduced the technique of intrasaccular wiring, in which coils of fine wire were fed into the aneurysm in the hope that fibrous tissue would form around the wire and fill the aneurysmal sac (Figure 1.2). The technique worked in its first clinical use, but the patient later died of sepsis.

Subsequently, many other physicians tried similar techniques to treat aneurysms, sometimes inserting iron wire, watch springs, or horsehair into the aneurysmal sac,



Figure 1.2 Specimen of sacciform aortic aneurysm removed after insertion of wire to promote thrombus formation and, thereby, to prevent rupture.

but rupture was not always prevented (Figure 1.3), and patients' life expectancies after these procedures could usually be measured in days, weeks, or months [12]. A particularly notable variant of this treatment method was tried by Duncan and Fraser, who in 1867 reported on their effort to obliterate a patient's thoracic aortic aneurysm by stimulating thrombosis within it with electrolysis delivered via a 13-cm needle. The aneurysm continued to expand after the treatment, however, and the patient died of hemorrhage 2 months later [13]. In 1879, Corradi combined Moore's intrasaccular wiring technique with electrolysis to create what became known as the Moore–Corradi method of electrothrombosis, variations of which were widely experimented with for many years afterward.

Open aortic surgery finally began to emerge in the early nineteenth century. In 1817, Sir Astley Cooper treated a 38-year-old man with a left external iliac aneurysm by placing a silk ligature on the abdominal aorta, which he had exposed via a transperitoneal incision - something Cooper had tried in a cadaver just two days earlier [14,15]. The patient lived for only 48 hours after the surgery, but Cooper's willingness to attempt such a difficult procedure impressed many of his colleagues, and he was eventually elected President of the College of Surgeons. (In his highly successful surgical practice, Cooper also became known for performing autopsies on his surgical patients whenever possible in order to learn from them.) Nonetheless, for the next 100 years, no patient would survive any attempt at aortic ligature. In 1902, Theodore Tuffier had brief success when he ligated the base of a saccular aneurysm of the aortic arch in an attempt to remove it, but ischemic necrosis developed and the patient died of hemorrhage two weeks later [16].

In 1888, American surgeon Rudolf Matas developed the concept of endoaneurysmorrhaphy: opening the

aneurysmal sac and using sutures to narrow the lumen from within, thereby removing the aneurysm while leaving blood flow intact [17,18]. Matas also had the idea of temporarily occluding large vessels during surgery to determine the consequences that permanent occlusion of these vessels might have; this test later became common surgical practice [19]. The findings from these occlusive tests made it clear that Matas' original procedure could not be used in the aorta or other major arteries without the risk of serious ischemic complications, so Matas developed a new endoaneurysmorrhaphy procedure in which the aneurysmal tissue was removed and a channel was created in the remaining, healthy tissue to allow blood flow [20]. This innovative procedure constituted a leap forward for aneurysm surgery, and a modified form of this technique is still commonly used today.

Nonetheless, before the twentieth century, most aneurysm surgeries ended in the death of the patient – if not from technical failures, then from post-operative infection. Marginally greater success was achieved in the middle of the last century with attempts to repair aneurysms by wrapping cellophane or other plastic films around the aneurysm to stimulate periarterial fibrosis and, thereby, to occlude the aneurysmal vessel (Figure 1.4). This method was first applied by Harrison and Chandy [21] to treat a subclavian artery aneurysm, and later by Poppe and De Oliviera [22] to repair syphilitic aneurysms of the thoracic aorta. These methods were successful in some instances, including the first successful repairs of blunt aortic arch injuries [23], but the outcome was too often unpredictable.

In 1951, while speaking at the annual meeting of the Southern Surgical Society, Denton Cooley and Michael DeBakey of Houston became the first surgeons to advocate the direct surgical removal of aortic aneurysms [24]. In the cases they presented, saccular aneurysms of the thoracic aorta, including the aortic arch, had been



Figure 1.3 Post-mortem specimen of aneurysm of ascending aorta, which ruptured despite extensive introduction of steel wire.



Figure 1.4 Treatment of an aneurysm by wrapping it in plastic film to induce fibrosis.



Figure 1.5 Drawings of three procedures performed by Cooley and DeBakey [24] in the early 1950s to repair aneurysms involving the aortic arch or its branches by clamping, excision, and aortic repair. (a) Repair of a subclavian artery aneurysm. a. = artery; car. = carotid; innom. = innominate; L. = left; Prox. = proximal; R. = right; subcl. = subclavian; sup. = superior; v. = vena/vein; vess. = vessel. (b) A completed repair of an aneurysm of the innominate artery and the adjacent portion of the aortic arch. Although the repair required the sacrifice of the right common carotid and subclavian arteries, the patient made a full recovery. (c) Repair of an aneurysm of the ascending aorta and the transverse arch. L. = left; n. = nerve; Sup. = superior. Reproduced with permission from [24].

successfully clamped, excised, and oversewn, so that the aneurysm was removed while aortic continuity was restored (Figure 1.5). In 1953, Bahnson [25] reported repairing several saccular aortic arch aneurysms, including one of traumatic origin. Impressively for the time, six of his eight patients survived.

The surgical repair of aortic coarctation also became a reality during the middle of the century. In the 1940s, surgical luminaries Alfred Blalock [26] and Robert Gross [27] each used animal models to develop techniques for the surgical repair of coarctation. These were put into practice in 1944 by Clarence Crafoord of the Karolinska Institute [28], who used end-to-end anastomosis to repair aortic coarctation in a 12-year-old boy. By 1956, Wright, Clagett, and colleagues had performed 10 coarctation repairs in adult patients [29].

The introduction of aortic grafts

Although the clamp-and-sew technique was very effective for treating localized sacciform aneurysms, these aneurysms were most commonly caused by tertiary syphilis, which, by the 1950s, was becoming increasingly rare. As a result, Fusiform and extensive sacciform aneurysms represented a greater proportion of the aortic aneurysms in need of treatment. Repairing these would require adequate graft materials to replace the substantial segments of the aorta that had to be excised.

In the early 1900s, Alexis Carrel and Charles Claude Guthrie [30,31] had conducted animal experiments in which homografts were used for aortic replacement. (This was part of the transplant research that would eventually win Carrel the Nobel Prize in medicine.) Gross and Hufnagel [27] had continued this line of research in the 1930s and 1940s in animal models of coarctation of the aorta, and Gross eventually used preserved homografts to repair aortic coarctation in human beings [32].

Graft repairs of aortic arch aneurysms were particularly challenging, partly because there were few reliable ways to prevent ischemic damage during the period of interrupted blood flow that such repairs required. Schafer and Hardin, in 1951 [33], were the first to attempt to use a homograft to repair an aneurysm of the aortic arch. The patient died of ventricular fibrillation immediately after the placement of 4 temporary polyethylene shunts intended to maintain cerebral and distal perfusion during the procedure. Two years later, Stranahan [34] repaired a syphilitic aortic arch aneurysm with a xenograft while a temporary shunt maintained blood flow between the ascending and descending aorta. The patient survived the procedure and had no apparent neurological deficit upon awakening but died shortly afterward from hemorrhagic complications of a left pneumonectomy that had been performed concomitantly with the aneurysm repair. In a similar procedure in 1955, Cooley and DeBakey [35] repaired an aortic arch aneurysm using prosthetic graft replacement and an ascending-to-descending aortic shunt, which included side branches to the carotid arteries. Nonetheless, even this shunting scheme did not prevent cerebral ischemia, and the patient died 6 days after the procedure. The next year, however, this Houston group successfully used a homograft to repair a fusiform aneurysm of the proximal aortic arch, which they did while the patient was on cardiopulmonary bypass (CPB) [36].

It eventually became apparent that homografts had limited life spans. Attempts were made to preserve graft tissue through freeze-drying, but the durability of such grafts was found to be highly variable [37]. Therefore, starting in the 1950s, many different synthetic materials were examined as potential alternatives, including nylon, Vinyon N[®] (a synthetic fiber made from polyvinyl chloride), Teflon[®], and Dacron[®] (polyester) [38–41]. It was found that nylon and Vinyon N deteriorated too rapidly, and Teflon, although durable, did not bond well with human tissue [42]. Dacron, therefore, became the material of choice.

The choice of fabric was not the only concern. Grafts made from fabrics whose weave was too porous had reduced durability and were associated with slower healing and increased risks of serious intra-operative bleeding and infection. This problem was addressed by weaving the material tightly and impregnating it with collagen, gelatin, fibrin, or similar substances to seal the interstices. In 1981, Cooley and colleagues reported another method of sealing woven Dacron grafts in which each graft was soaked in the patient's own plasma and then placed in a steam autoclave, thereby filling the interstices of the graft material with coagulated protein [43]. This measure substantially reduced post-operative mortality and bleed-ing complications [44], and it inspired many subsequent improvements in commercially manufactured grafts.

Protection against ischemic injury

Of equal importance as the development of graft materials to the evolution of aortic arch surgery was the introduction of measures to protect the central nervous system and the vital organs against ischemic injury. Temporary shunts could be used in some cases, but doing so added considerable time to the procedure, and, as noted above, the shunts did not always provide adequate protection. Additionally, surgery on aneurysms (especially fusiform aneurysms) in critical parts of the aorta, including the transverse arch, required temporary circulatory arrest while the repair was completed. Therefore, preventing this type of injury during cardiac, coronary, or aortic surgery would require some means of perfusing the vital organs and of reducing their metabolic requirements.

Cardiopulmonary bypass

In the early years of cardiovascular surgery, some clinicians experimented with cross-circulation, in which the heart and lungs of a 'donor' would circulate and oxygenate the patient's blood while the patient's own heart and lungs were disconnected from the circulation [45]. This cumbersome and potentially hazardous technique was abandoned after the introduction of effective mechanical pump oxygenators. The first of these devices was designed by John Gibbon of Jefferson Medical College. After more than a decade of work on the device, Gibbon put it to the clinical test in 1953, using it to support 4 patients during open-heart procedures to repair congenital cardiac defects [46]. Only 1 patient survived, however, and Gibbon abandoned his work on the pump. The device was later simplified and improved upon by DeWall and Lillehei [47], who added a bubble oxygenation system with a defoaming coil to return the blood to a purely liquid state.

With the advent of CPB came considerable debate about cannulation schemes and direction of blood flow. Crawford and colleagues [48] used antegrade perfusion in an aortic arch repair, a technique that came to be commonly used in the 1960s and thereafter. Retrograde cerebral perfusion through the superior vena cava, first used by Mills and Ochsner [49] in 1980 to treat a massive air embolism during CPB, began to be used in aortic aneurysm repair shortly thereafter. The continuous retrograde perfusion technique subsequently developed by Ueda [50] for aortic arch procedures is still in use today. Now anterograde cerebral perfusion through the aortic arch vessels is also increasingly popular; this technique was revised for use in aortic arch replacement by Frist in 1986 [51]. Additionally, CPB is not used for distal arch repairs; crossclamping of aneurysms that begin in the distal aortic arch and that do not involve the innominate artery can be accomplished safely without bypass or shunting if the repair is accomplished quickly and efficiently [52]. Autotransfusion techniques have enhanced such procedures.

Hypothermia

Together with CPB, induced hypothermia and total circulatory arrest made it possible to repair aneurysms in any region of the aorta. The notion of using hypothermia to slow the metabolism of the brain to increase its ischemic tolerance during open cardiovascular procedures was introduced by Wilfred Bigelow of Toronto General Hospital, who examined the effects of surface cooling in animal models of cardiac surgery [53]. The first successful clinical use of Bigelow's cooling technique was made by Lewis and Taufic [54] at the University of Minnesota in the repair of an atrial septal defect in a 5-year-old girl. Bigelow also discovered that barbiturate administration provided cerebral protection during hypothermia, a finding that was later confirmed in clinical studies [55].

While Bigelow was performing his surface cooling experiments in Toronto, Ite Boerema and colleagues in Amsterdam were experimenting with central cooling and re-warming [56], in which blood was removed from an artery, cooled or warmed by an external device, and then returned through a vein. This work led to the development of cooling methods that could induce deep hypothermia (i.e. cooling to approximately 10°C). The combination of deep hypothermia and circulatory arrest with open anastomosis was first used to treat extensive aortic arch aneurysms by Christiaan Barnard in 1963 [57]. It was subsequently used by Dumanian [58] to repair a traumatic aneurysm of the transverse arch in 1970, and for prosthetic replacement of the aortic arch by Griepp [59] and by Ott and Cooley [60]. Deep hypothermia and circulatory arrest provided considerable protection of the central nervous system during the procedure, but the technique was not without disadvantages; it was time-consuming, and it could cause coagulopathy, which increased the patient's risk of intra-operative bleeding and post-operative stroke and death [61]. As a result, in 1981, Cooley, Livesay, and colleagues recommended initiating moderate systemic hypothermia and shortened periods of total circulatory arrest after the aortic arch vessels were clamped [44].

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Several advances in the conduct of aortic arch repair have been made in recent decades, many of which resulted from efforts to simplify procedures. For example, the introduction of 'open' distal anastomosis, in which only the proximal end of the aneurysmal aortic segment is clamped, has increased the speed with which grafts can be anastomosed to the aortic arch and the aortic arch vessels reimplanted [44]. Together with the introduction of biological glues by Bachet [62] and others, this technique has reduced operative mortality in aortic arch surgery. A similarly useful simplification of technique for the repair of aneurysms involving both the arch and the descending thoracic aorta was Borst's two-staged 'elephant trunk' procedure, in which the diseased arch segment is repaired first, and the distal end of the vascular graft is left inside the descending segment, thereby simplifying and rendering less invasive the creation of the distal anastomosis in the second stage of the procedure [63]. For patients whose aneurysms are larger in the descending or thoracoabdominal portion of the aorta than in the arch, Carrel and Althaus developed the 'reversed' elephanttrunk procedure, in which the graft is placed in the proximal descending aorta and folded in on itself during the first stage of the operation. The folded portion can then be pulled out with a nerve hook and used to replace the transverse aortic arch during the second stage of the procedure [64]. This technique is very useful for aneurysms that involve a large portion of the aorta [65,66].

For aneurysms that extend into the arch from the aortic root and involve annuloaortic ectasia, Bentall developed a procedure for replacing the entire diseased segment and aortic valve with a fabric graft containing a mechanical prosthetic valve [67]. This procedure remained standard of care for 25 years, but like all mechanical valve implantations, it required the patient to take anticoagulant medications indefinitely after the procedure. Therefore, in the early 1990s, Yacoub [68] and David [69] each devised alternative procedures in which the native valve could be spared by reshaping the aortic annulus (Yacoub) or by mobilizing the native valve and reimplanting it inside the synthetic graft (David).

Combined surgical and endovascular approaches

Simultaneous with the refinement of surgical approaches to aortic arch repair has been the rise of modern endovascular ones. Although endovascular stent-grafting has been used successfully in the abdominal aorta and, more recently, in the descending thoracic aorta, strictly endovascular repair of aortic arch aneurysms poses particularly difficult technical challenges. The curve of the arch complicates stent deployment in some cases, and, more importantly, deploying stents in the arch can occlude one or more of its branch vessels. This occlusion may be tolerable in the left subclavian artery [70,71] (unless the aneurysm involves this artery) but not in the left common carotid or innominate arteries.

For these reasons, hybrid procedures have begun to be developed for aortic arch repair. These are generally 2-stage procedures in which open surgery is performed first to create landing zones for the graft [72], to transpose or revascularize aortic arch vessels to prevent ischemic complications after stent-graft placement [73,74], or to insert the stent-graft in elephant-trunk fashion (after which the distal end of the graft is secured during the second stage of the procedure) [72,75]. These procedures



Figure 1.6 Drawing depicting the objective of curative surgery for total aortic aneurysm with restoration of vascular continuity using a fabric graft. Current treatment may involve endovascular techniques with covered stents. Reprinted from [76] with permission from Elsevier.

have produced good short-term results, but little is yet known about their long-term outcomes.

Conclusions

Human beings have been aware of aneurysms for millennia, but only in the past century has surgical repair of the aortic arch progressed from being impossible to being a desperate last resort to becoming a viable treatment option. Aortic surgery has become sufficiently sophisticated that, using CPB and other adjuncts, it is now possible to replace the entire vessel, from the aortic annulus to the bifurcation, with a synthetic graft (Figure 1.6). Today, the objective of any arch repair procedure is not merely to remove the aneurysm but to restore circulation to all vital tributaries. Further improvements in surgical adjuncts and in hybrid surgical/endovascular techniques will make this goal achievable in an ever larger proportion of patients than is possible today.

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