Clinical Research

Aortic Diseases in India and Their Management: An Experience from Two Large Centers in South India

Introduction
Aortic diseases are to a great extent a disease of the elderly and are becoming more common as the population ages. They account for a significant cardiovascular morbidity and mortality worldwide. The incidence of aortic diseases in India is expected to rise with the increasing age of the population. Diagnostic evaluation of aortic disorders has improved in the last two decades, allowing earlier diagnosis and therapeutic intervention. We summarize the major disease entities affecting the aorta in India and review their current management in two large centers in South India.

Current Practice
In India, atherosclerosis constitutes the principal etiopathogenesis for aortic diseases that are classified into chronic disease states and acute aortic syndromes.

Chronic disease states
- Congenital – coarctation of aorta
- Degenerative – aortic aneurysm and aorto-iliac occlusive diseases
- Connective tissue disorders – aortic dissection (AD) and aneurysm
- Inflammatory – aortoarteritis, aortic aneurysm, and aorto-arterial steno-occlusive disease.

Acute aortic syndromes
- Ruptured aortic aneurysm
- Acute AD
- Penetrating atherosclerotic ulcer (PAU)
- Intramural hematoma (IMH).

Of the numerous clinical conditions, abdominal aortic aneurysm (AAA) constitutes the most common of aortic diseases forming the flagship clinical entity that has been the prototype around which nearly all refinements in diagnosis and treatment strategies occurred in vascular surgery.

Although coronary artery and extracranial carotid artery diseases are prevalent in India like in the developed countries, aortic diseases appear to be of much less incidence compared to developed countries as per the number of patients treated by vascular surgeons across our country. Longer life expectancy, dietary habits, and Caucasian ethnicity may be the predisposing factors for higher prevalence in Western world.

This chapter will mainly focus on aortic aneurysmal disease in detail, mentioning the other clinical disease entities in brief, as practiced between the two largest centers of aortic surgery in India where the authors come from.

Abdominal Aortic Aneurysm

Clinical evaluation
1. Presentation: Unlike elsewhere in the world, a good subset of patients (50%) present with symptoms referable to AAAs (and likewise thoracic). Only 10% of patients present with rupture in abdominal aortic domain and another 40% detected incidentally while being evaluating for unrelated causes.
2. Age group: Most patients belong to 55–65 years group, with only 10% presenting beyond 75 years, having male preponderance, with 9:1 male to female ratio. Comorbidities are usual with high incidence of hypertension, chronic obstructive pulmonary disease, and smoking.
3. Investigations: Ultrasonography is the initial preferred diagnostic modality. Specific investigation such as computerized tomography (CT) aortogram of the abdomen including lower chest is performed in every single patient for delineation of surgical anatomy for the last three decades. Whenever renal dysfunction is detected or ruptured aneurysm is suspected by the classic clinical triad, contrast injection is avoided, utilizing ultrasonography and noncontrast CT scan for AAA.
4. Complete hematologic and biochemical investigations, coronary work up, and respiratory evaluation are mandatory for all patients with aortic aneurysmal disease.
5. Threshold for AAA repair is ≥5.5 cm and thoracic aneurysm is ≥6 m. In female patients, ≥5 cm is taken as threshold, although a number of afflicted and treated patients is small in number.[1,2]

In the studies of abdominal aortic size in CT scan performed for vascular and nonvascular indications and based on body surface area calculated using Mosteller formula from individual patient’s weight in kilograms and height in meter,[3] a personalized threshold is derived, helping to choose the appropriate size and time to decide intervention in asymptomatic, the so-called small AAA patients measuring 4.0–5.4 cm.

Surgery
- Open surgical repair is performed in standard fashion using transperitoneal midline xiphopubic laparotomy in almost all patients. Endovascular repair is performed only on selective indications. From 1998 till date, all 716 patients have been treated for AAA, of which 554 (77.4%) patients (467 [84.3%] elective and 87 [15.7%] ruptured) have been surgically operated upon by the authors. Coated polyester straight tube...
Whenever possible, large graft inclusion repair could be achieved in 70% of patients and rest required uni/bilateral iliac aneurysm repair mandating bifurcated aortic prosthesis. Sri Chitra Institute uses albumin/gelatin-coated Dacron grafts whereas the Narayana group uses a variety of grafts including ePTFE (Gore-Tex, Atrium) and gelatin- and albumin-coated Dacron grafts (Maquet, Hemashield, and Vascutek). Elective open surgery mortality is currently 3.2% and 25% in ruptured AAA patients which justifies our heavy bias toward open repair then and now.[6] Our patients arrive late due to poor transportation and are mostly hemodynamically unstable on presentation. In the same period, 162 (22.6%) endovascular aneurysm repairs (EVARs) (144 [88.9%] elective and 18 [11.1%] ruptured) were performed. In EVAR group, the secondary intervention was necessary in 3.08%. Elective EVAR mortality is currently 0.69% and 27.7% for ruptured AAA patients. There were 11 (6.8%) endoleaks: Type I (9.1%), Type II (81.8%) (stable 7/9 (77.8%); re-intervened 2/9 (22.2%), and Type III (9.1%). There were two limb occlusions treated by femoro-femoral crossover bypass.

- During the last 4 years, 18 patients with juxtarenal AAA underwent open surgery. A modified top-end strategy, essentially a triad of surgical adjuvants, namely, (a) division and suture closure of the left renal vein flush with inferior vena cava, (b) inter-renal aortic clamping maintaining visceral and upper renal artery perfusion, and (c) renal preservation fluid through the lower renal artery during proximal anastomosis. One patient required right renal artery stenting for acute renal failure subsequent to snow ploughing effect into the right renal artery ostium following inter-renal aortic cross clamp.[8] Seven (8.6%) patients had a transient rise in creatinine, of which one patient required temporary dialysis.

**Thoracic Aneurysms**

Vascular surgeon's domain starts from aortic arch moving across to descending thoracic aorta (DTA) followed by thoraco-AAA (TAAA). Till 2007, open repair was resorted to all patients needing intervention. However thereafter, aortic stent grafting became the frontline therapy for thoracic domain except in selected patients in good health, aged <60 years and large aneurysms presenting with dysphagia.

At Present, TAAA forms the main indication for open repair. Open repair of TAAA is performed using thoraco-phreno-retroperitoneal approach to reconstruct the aneurysm and additional upper thoracotomy in case of Crawford Type I and II extent, with inclusion graft technique, re-implanting viscero-renal arteries and intercostal arteries.[4] Whenever possible, large intercostal arteries are included in beveled proximal/distal aortic anastomosis and smaller ones are sutured off. Cerebrospinal fluid drainage is employed in all patients since 2005 onward. Distal aortic perfusion is achieved using temporary aorto-femoral bypass, from where blood is siphoned out to provide perfusion to viscero-renal arteries using Pruitt-Inahara® shunts. Cold renal preservation solution (Custodiol®) is used in most of the procedures before transoastelial blood perfusion.

Vascular registry documented 168 patients with TAAA having undergone elective conventional open repair. There were Crawford 29 (16.4%) Type I, 42 (25.0%) Type II, 7 (4.2%) Type III, 86 (51.2%) Type IV, and 4 (2.4%) Type V TAAAs.

There was an overall mortality of 35/168 (20.8%) and all survivors discharged to their homes, without needing to enter into the secondary high-dependency centers (for spinal cord or renal dysfunction). Thirty percentage of patients had Takayasu’s arteritis as etiopathogenesis for TAAA in our series. Poor outcomes were recorded in patients with prior ischemic heart disease, respiratory dysfunction, prior abdominal surgery, and extent of surgery.

**Coarctation of Aorta**

Patients with coarctation of aorta in adulthood, in particular, present with severe/intractable cephalocervicbral hypertension. Primary choice at the Sri Chitra Institute is open repair by excision of coarctation segment followed by reconstruction of aorta in the majority with interposition prosthetic graft.[5] In a small subset, graft bypass from the left subclavian artery to DTA was also chosen due to technical reasons. We have performed 87 adult coarctation reconstructions without mortality or major morbidity.[6] The Narayana Institute utilizes endovascular technique preferentially in coarctation of the aorta. One hundred and six procedures have been performed with no mortality. Covered stents were used in 40% of cases.

**Aorto-Iliac Occlusive Disease and Middle Aortic Syndrome**

Former as a result of atherosclerosis and latter as Takayasu’s disease are dealt with balloon aortoplasty in the majority of patients. Flush aortic occlusions are addressed by open surgical approach. Aortic thromboendarterectomy is facilitated by supraceliac aortic cross clamping, vertical aortotomy 5 mm below the lower renal artery allowing disobliteration of the aorta in about 10 min, and aortic graft anastomosis performed after shifting the aortic cross clamp to infrarenal aortic position. Aortobiemoral bypass graft procedure was accomplished in usual fashion.

**Aortic Stent Graft Procedure**

Beyond 2007 till date, 110 endovascular aortic repairs were performed; of which EVAR was only 10 in number in selected AAA patients. Hybrid aortic arch, for degenerative
Aortic Dissection

Stanford-A AD with its intimomedial tear and intimal flap commencing in the supracoronary ascending aorta is beyond the scope of vascular surgeon's domain. Stanford-B AD with intimomedial tear starting in postsubclavian DTA is initially treated in the line of modified Wheat and Palmer regimen.[7] In complicated cases, early TEVAR with or without fenestration or covered stent to treat malperfusion was used in four patients out of the 57. In eight patients above >75 years and having high comorbidity index, medical management was pursued. Eleven patients underwent open surgical repair using cardiopulmonary bypass and deep hypothermic circulatory arrest with nine survivors. Totally, 53 patients underwent standard/hybrid TEVAR as dictated by the close proximity of intimomedial tear to the left subclavian artery origin. Mortality rate was 4/54 (7.0%). One patient succumbed to visceral malperfusion in the perioperative period, the second patient expired during the conversion to open repair at 18 months following TEVAR due to aneurysm dilatation of dissected aorta, and two patients died during follow-up.

Acute Aortic Syndromes

Four in all, acute aortic syndromes cause severe distress and a subset mandates immediate attention (ruptured aortic aneurysm and Stanford-B AD with complication) and others at planned timeline.

Having had poor results with ruptured TAAA, no surgical option is given to these patients. In contradistinction, during the last 5 years, 57 AAA patients presented with rupture, for which our only option was open surgery leading to 75% early survival. In our setting, we do not offer EVAR in rupture instance. We have had only a few patients with IMH and PAU, who were given open/endovascular repair.

Primary Aortic Mural Thrombosis

Primary aortic mural thrombus (PAMT) is an uncommon condition but important source of noncardiogenic emboli with a difficult diagnosis and a high rate of complications including high mortality. We reported our experience (largest in the world) of thrombo-embolic disease from PAMT and reviewed its contemporary management. A total of 88 patients presented with acute occlusion of the extremities or visceral arteries between January 2011 and September 2013 were included in this study. Of these, 19 patients (mean age 41.2 years; male: female ratio 1.2:1) had a major thrombotic or embolic source within an otherwise normal aorta after thorough evaluation of heart and great vessels. In ten patients, thrombus was located in the thoracic aorta, three in the perivisceral abdominal aorta, and six in the infra-renal aorta. Thrombus in thoracic aorta was treated with stent grafts in four patients, bare metal stents in three patients, and anticoagulation alone in two patients. In suprarenal abdominal aorta, all three patients underwent trapdoor aortic thrombectomy. Infra renal aortic thrombus was managed by aorto-bifemoral embolectomy in two patients, aortic stenting in two patients, surgical thrombectomy in one patient, and oral anticoagulation alone in one patient.

Successful treatment was defined as freedom from further embolic events or recurrence of thrombus achieved in (14/19) 76.4% patients with a mean follow-up period of 16.2 months (range 2–28 months). There were 4 (21%) thrombus-related deaths, all due to primary thromboembolic insults. One patient needed a below knee amputation due to recurrent thrombotic episode.

Symptomatic PAMT is an uncommon but important source of noncardiogenic embolus. It appears to occur more frequently in young female adults. Endovascular coverage of the aortic thrombus, when feasible, appears to be an effective and safe procedure using either stent grafts or closed cell metal stents. When a thrombus is located adjacent to visceral vessels, it should be managed with an open trapdoor thromboembolectomy.[8]

Summary

Our experience at two major institutes in India reflects aortic practice in our country. There is no National Vascular Registry available as of now.

In India, currently, EVAR have largely broadened the therapeutic strategy for aortic aneurysm. It is estimated that on an average, 500 procedures being performed by vascular surgeons, interventional radiologists, and cardiologists, of which 150 open surgery and 350 endovascular aortic repairs annually.

Taking a leaf from the worldwide burden of surveillance, the need for secondary re-interventions, a small but distinct (continued) risk of rupture of aneurysms treated by endovascular repair and availability and affordability of endovascular repairs, we have in place a robust and perseverant protocol of conventional open surgery for AAA and selective EVAR for elderly patients reporting with high comorbidity index, hostile abdomen, or stable hemodynamics allowing procurement of devices and planning repair.

In view of the fact that our patient's body surface area is between 1.4 and 1.8 m², accepted threshold to intervene is ≥5.5, which may not be suitable for intervention/surgery in
India. We have worked out a personalized threshold to decide optimal size and time to intervene, as we believe personalized and not universal threshold is pertinent to our patients.

A triad of surgical adjuvants described earlier has helped to treat juxtarenal AAA successfully although only in 18 patients recently. These adjuvants are already of proven value individually, but three components judiciously used during surgery have benefitted the complex subgroup of juxtarenal AAA with excellent recovery and quality of life.

Endovascular aortic repair has evolved as a primary therapeutic strategy across India, wherein paradigm shift to perform aortic stent grafting for aneurysm and Stanford-B AD in varying proportion exists today. Although numbers are small, the vascular practice has kept up with changing trends and strategies across the country with excellent results. However, we believe in a country like India with limited resources and access to endovascular surgery, AAA seems to be a domain for open surgery with its proven durability and easy surveillance with EVAR being used in selected patients. Although data from the recent trials show the early survival benefit with EVAR compared to open repair at 1 month, survival at 2 years equalizes with open and EVAR groups for AAA repair.

In thoracic aortic domain, in contradistinction, endovascular repair is going to be the therapy of choice for all thoracic aortic pathologies for the obvious reason of early and late success in most institutions worldwide.

**Madathipat Unnikrishnan, Ajay Savlania, Prakash Goura, Himanshu Verma, Ramesh K Tripathi**

Department of Cardiovascular and Thoracic Surgery, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram, Kerala, Department of Vascular Surgery, Narayana Institute of Cardiac Sciences, Bengaluru, Karnataka, India

**Address for correspondence:** Prof. Ramesh K Tripathi, E-mail: ramesh.tripathi@vascularsurgeon.org

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