ASCENDING AORTIC ANEURYSM, WANDERING TOWARDS OLD PROBLEM WITH NEW INSIGHT

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ABSTRACT

Ascending aortic aneurysm is a silent and highly lethal disease. Generally, ascending aorta grows asymptomatically until it dissect or rupture. In case of acute complication mortality is as high as 90%, whereas if treated on time mortality and morbidity decreases significantly. Pathophysiology of the Ascending aortic aneurysm is a complex process. Aorta itself is an organ and it should not be considered merely as a tube transporting blood from heart to the organs. Several non-invasive and invasive imaging methods are available at disposition to diagnose this indolent killer at early stage. However, discrepancies exists about when to operate these patients. Several other methods has been reported to identify these patients at risk of dissection or rupture. Conservative treatment could be used in patients with small aortic diameter, but effectiveness of such approach is under scrutiny. Emerging endovascular treatment using stents should be advocated with caution. Surgical treatment, the gold standard, is recommended for asymptomatic patients with aortic diameter of 5.5cm, whereas 4-4.5cm for patients with Marfan's disease. Postoperative morbidity and mortality has significantly decreased due to better anesthetic management, improved surgical techniques and progress in preoperative and postoperative care.

KEYWORDS : Ascending aortic aneurysm

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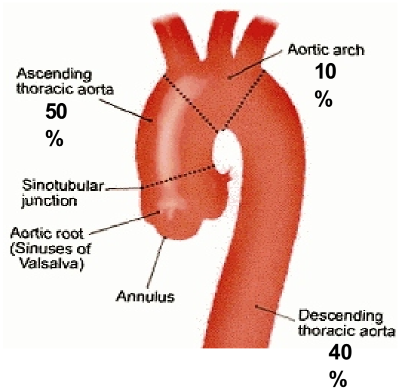
BACKGROUND
Epidemiology

Cardiovascular diseases are one of the most important cause of death in the majority of developed countries. Cardiovascular morbidity and mortality are in rise in many developing countries. Among cardiovascular pathologies, aortic aneurysms probably represent the most lethal and indolent enemy of the medical community. Usually, they silently and asymptomatically grow up until an acute and often catastrophic complication occurs. The threats of a non-operated aortic aneurysm include dissection or rupture of the aorta, subsequently leading to death. In contrast, despite the armamentarium of modern perioperative and post-operative cardiac surgical care, the risk of surgery include paraplegia, stroke, bleeding, and mortality ranging from 3 to 9% after elective surgery. Conversely, in cases of acute events mortality can be as high as 90%. In the United States, aortic aneurysms (thoracic and abdominal) constitute the 17th leading cause of death in the general population and the 15th for individuals older than 65 years. Approximately 15,000 individuals die every year from this pathology in the United States of America', which is more than death caused by HIV infection. Even if less frequent than abdominal aneurysm, thoracic aortic aneurysms (TAAs) have a reported incidence of about 10 cases per 100,000 patients/year, with a yearly risk of rupture or dissection of 7%. Significant heterogeneity occurs in distribution of aneurysm disease along the aorta, with 50% of all TAA involving the ascending aorta, 10% the arch, and 40% the descending aorta[2,3] (Figure 1).

Cumulative and yearly risks of aortic dissection and rupture increase as the aneurysm size increases. Incidence of dissection in female patients have been shown to be higher than male, which could be due to a given aortic diameter representing proportionally greater dimension in smaller women. In front of a similar distribution of TAA between sexes, 79% of acute complications inexplicably occur in women''.

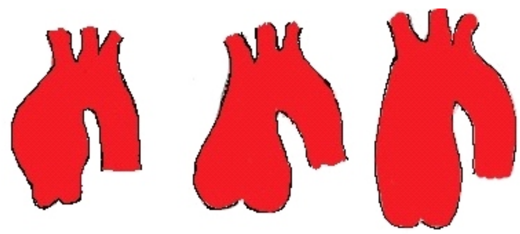
Figure 1: Anatomic localization of TAA



Definition

Aortic aneurysm can be defined as a localized undue dilation with 50% increase in size over the normal diameter. The "normal diameter" strictly depends on age, sex and body size, as well as the anatomical localization of the affected aorta. For the physiologically smaller abdominal aorta, the term aneurysm is usually limited to diameters exceeding 30 mm, while, on the contrary, a TAA should be conventionally larger than 40mm. According to Elefteriades J, ascending aortic aneurysm are divided into three categories, according to the pattern of the involvement of the aortic root. These are supracoronay aneurysm, annuloaortic ectasia (Marfanoid) and tubular diffuse enlargement of the aorta. (Figure 2)

Figure 2: Types of ascending aortic aneurysms



Symptoms

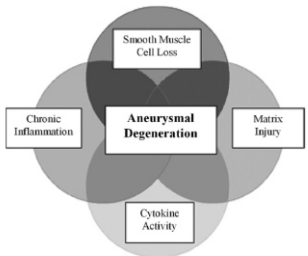
Patients with a TAA are usually asymptomatic and diagnosed by chest X-rays or CT-scan requested for other reason. Thoracic aortic aneurysm can cause symptoms by compressing nearby structure. Hoarseness of voice could be a presenting symptom if it compresses recurrent laryngeal nerve; stridor, dyspnea from tracheal, bronchial or lung compression; dysphagia from esophaseal compression; and plethora and edema, from superior vena cava compression. A dull and vague pain in neck and jaw may be an indication of aortic arch aneurysms, while back, interscapular, and/or left shoulder pain may occur with descending aortic aneurysms. dissects or ruptures. Frequently, patients present with sign and symptoms of aortic insufficiency, which ought to be the main reason behind patient coming to clinical attention. Ultimately, acute syndrome including aortic dissection or aortic rupture, if could make to the hospital, might present with potentially lethal outcomes.

Pathophysiology

Pathophysiology of ascending aortic aneurysm is complex. Risk factors for the development of thoracic aortic aneurysms include hypertension, smoking, and chronic obstructive pulmonary diseases. Ascending aortic aneurysms are also related with bicuspid aortic valve. Additionally, several genetic syndromes with a predisposition for ascending aortic aneurysms have been identified. Most common genetic diseases effecting thoracic aorta are: Marfan Syndrome, Loeys-Dietz Syndrome, Ehler-Danlos Syndrome, Turner Syndrome, familial thoracic aortic aneurysm Syndrome, and still others are idiopathic.

Historically, it was supposed that aortic dilation and strength reduction of the aneurysmatic aortic wall were exclusively related to primary connective tissue alterations. Recent data support that aortic wall remodeling is a dynamic process with active involvement of the immunological system and smooth muscle cells (SMCs). TAA formation and progression is a multifactorial process that involves both cellular and extracellular Mechanism.

Figure 3: **The pathogenesis of TAA basically involves four essential mechanisms**



- ♦extracellular matrix proteolysis
- ♦chronic inflammation
- ♦cytokine activity
- ♦smooth muscle cell (SMC) loss

These four mechanisms interact to undermine the integrity of the aortic wall leading to aneurysm formation, growth, and, ultimately dissection or rupture.

Histopathology

Pathologically, cystic medial necrosis and lamellar medial necrosis are found in the arterial wall of the dissected aorta with helicoids hypertrophy of the collagenous fibers, rarefaction and fragmentation of the elastic fibers in the tunica media. It has shown that there are abnormalities in collagen and elastin in the tunica media of the dissected aorta. The pathological findings of patients without Marfan syndrome show rupture and decrease in elastic fibers in the tunica media. Thus, it is considered that fragility of the tunica media is still a cause of aortic dissection in patients without Marfan syndrome. According to Illiopoulos and co-authors the ascending aortic tissue undergoes major wall remodeling on diameter enlargement, with the decrease in media partly compensated by an increase in intimal thickness. They have also found that elastin and collagen area densities decreased in the media layer of TAA specimens as compared to control specimens. Furthermore, they have reported a decreased elastin density in the intima and entire wall of TAA and an increased collagen density in the adventitia and an invariant density in the entire wall. Decrease elastin content substantiate the deficient extensibility of ascending aortic aneurysm.

The extracellular matrix (ECM), which is made of the same elastic fibers and collagenous fibers that exist in the tunica media, plays a role in maintaining the shape of the aortic wall. The matrix metalloproteinases (MMP) are a heterogenous family of enzymes that, altogether, are capable of degrading all the constituents of the ECM. They are required for normal development and for general turnover of the ECM, but are elevated in many pathological conditions including inflammatory diseases. MMPs are tightly regulated at three main points, transcription, activation, and inhibition. All of them are secreted in a proform, which requires initial activation by another proteinase, and once activated, there are at least four specific MMP inhibitors (tissue inhibitors of Metalloproteinase, or TIMPS), which can bind them irreversibly.

The role of MMP-2 (Gelatinase A) and MMP-9 (Gelatinase B) in the development of aortic aneurysm has been extensively studied. MMP-2 and MMP-9 generated in the macrophages cut the type IV collagen in the elastic fibers and basement membrane, and break the tunica media of the aortic wall, resulting into a fragile and enlarged aorta. MMP-9 is also produced by fibroblasts or SMCs that have been stimulated to undergo a phenotypic switch favouring synthetic activity. Most of the studies showed the role of MMP-2 in abdominal aortic aneurysm, whereas the contribution of MMP-2 to the development of TAA is not fully defined. Studies focusing on idiopathic TAAs have failed to show an increase in MMP-2 levels compared to normal aortas.

MMPs degrade the structural proteins of the aortic wall. These enzymes are normally antagonized by TIMPs and fibrillin-1 gene. Recently, it has been demonstrated an excessive lysis of matrix in ascending aortic aneurysms and dissections. It is hypothesized that aneurysm patients are genetically prone to excessive MMPs activity, leading ultimately to degradation and thinning of the aortic wall with subsequent increase of the aortic diameter and aneurysm formation.

Inflammatory and immune mechanism

Inflammatory cells like monocytes, macrophages, plasma cells, B-lymphocytes, and T-lymphocytes actively participate in the genesis of aneurysms. Tang et al. suggested that the outward vascular remodeling and intimal expansion of TAA correlate with the extent of a Th1 immune response ("IFN-γ driven")[11], which causes production of cytokines such as IFN-γ, TNF-α and various chemokines, all contributing to leukocyte recruitment, cell-mediated inflammation and tissue damage.

Biomechanical property of the aorta

Result of above mentioned pathophysiological processes lead to alteration of one of the important property of the aorta, i.e. biomechanical property. Biomechanical properties of the aorta may have additional value regarding the occurrence of aortic dilatation, dissection, and rupture in aortic degenerative disease and may contribute to the risk stratification of patients at risk for aortic complications. The ultimate goal in the care of a patient with an aortic aneurysm is to prevent rupture, a material failure that occurs when the aortic wall stress exceeds the tensile strength. According to the law of LaPlace, wall tension is influenced by intramural pressure, vessel diameter, and wall thickness, and the application of this tension to a defined area indicates wall stress. Therefore, the aorta, we are realizing, is "much more than a tube", as stated by a pioneer in the study of ascending aortic aneurysm pathology. It is more complex than passive, hollow conduit for delivery of blood to the vital organs. Aorta is rather to be considered as an organ itself, with complex histological structure and intrinsic biology, which dynamically interact to create a sophisticated mechanical bio-machine that works to optimize ventricular work and control delivery of blood and signals to tissues. The material properties of the normal and aneurismal aorta with regard to breaking stress and distensibility or stiffness have been an area of intense study.

Once aneurysm formation is initiated, it is associated with destructive remodeling of the aortic wall, the time course of which is characteristic of steady structural deterioration, radial enlargement, rearrangement of hemodynamic loads, and ultimately rupture. The biomechanical behavior of arterial tissue is generally attributed to the status of the structural proteins present. Elastin provides distensibility and recoil at lower pressures, whereas collagen provides tensile strength and stiffness at higher pressures. Loss of elastic fibers and derangements in collagen cross-linking seems to be involved in the TAA. Aneurysm rupture is a biomechanical failure that occurs when the stresses exerted on the aortic wall by hemodynamic loads exceed the tissue's capacity to sustain stress. Knowledge of the mechanical properties of aortic aneurysms, together with their individual geometry, is vital in predicting their risk of rupture. Biomechanical properties of the aorta can be expressed in terms of distensibility (percentage of luminal increase per pressure increase {in millimeters of mercury}during a heart beat cycle) or the propagation velocity of the pulse or flow wave through the aorta (pulse wave velocity, flow wave velocity propagation)". Koullias and colleagues offered an estimate of the *in vivo* mechanical properties like distensibility, aortic wall stress and Incremental elastic modulus of TAA. Okamoto and associates examined the *in vitro* properties (opening angle, biaxial elastic, and uniaxial circumferential strength tests) of dilated ascending aorta, and Vorp and coworkers compared the biomechanical properties (Maximum tissue stiffness and tensile strength) of TAA with respect to the non-aneurismal aorta, reporting lower strength and higher stiffness on the former. A change in aortic radius caused by an incremental change in pressure describes vessel distensibility, and a lack of distensibility or resistance to deformation is referred to as stiffness. Aortic distensibility decreases with increase in aortic diameter, which emphasize aortic stiffness as a potential predictor of aortic dilatation and aortic dissection in Marfan patients. Several studies evaluated by echocardiography and MRI have demonstrated increased aortic stiffness in TAA patients'.

There has been strong evidence from many reports that the mechanical properties and strength of aortic wall tissue show pronounced age dependency. In particular, Okamoto and colleagues reported a negative correlation of failure stress with age for the dilated ascending thoracic aorta, whereas same finding for the healthy descending aorta has been demonstrated in several other studies[81,89,]. Other studies also disclosed that the aorta becomes less extensible and stiffer as a result of aging.

DIAGNOSIS

Instrumental evaluation of the ascending aorta

For the diagnosis of aortic aneurysm several diagnostic tools are available ranging from non-invasive examination (Chest X-ray, Echocardiography, Computed tomography and MRI) to invasive examination like angiography. Noninvasive imaging is essential for assessment of aortic size and in some cases functional parameters. It is important to know the accurate size of the aorta because key decisions regarding management of the aortic aneurysm depend on size.

Chest X-Ray

Chest X-ray often performed as a part of general examination in patients with potential cardio-pulmonary disease. It sporadically detects abnormalities of aortic contour or size that require definitive aortic imaging.

Echocardiography

Echocardiography is one of the most used imaging modality in the cardiology, which has a high sensibility and specificity in diagnosing variety of cardiac pathology including ascending aortic aneurysm. Transthoracic echocardiography (TTE) is more readily available, easy to use, transportable and cost effective. No need of contrast and sedation make it as a first line diagnostic tool in clinical set up. However, transesophageal echocardiography (TEE) is superior to TTE and more accurate for assessment of the thoracic aorta, but sometime requires sedation and has a small risk of complications like esophageal perforation (less than 0.03%). In addition, with diagnosis of aortic dilatation, echocardiography may reveal other associated pathology that suggests the underlying etiology of the aortic disease (eg, bicuspid aortic valve). Nevertheless, for accurate evaluation of ascending aortic aneurysm and to confirm the indication for surgery, there are some restrictions. TTE and TEE are user dependent. TTE can only visualize the proximal part of the ascending aorta, thus it can miss an aneurysm of the mid-portion of the ascending aorta. Even TEE is limited by the interposed tracheal air column and can be "blinded" to the upper portion of the ascending aorta. Furthermore, there is no universal agreement for exact place of aortic diameter measurement and whether the aortic wall should be included or excluded in the aortic diameter measurement.

CT Scan

New generation helical CT scan has a sensitivity up to 100% and specificities of 98% to 99% for diagnosing abnormalities of the thoracic aorta. IRAD data shows that in some centers, CT has been used more frequently than Echo in case of aortic dissection. Major advantages of CT scan are: very wide availability, ability to image entire aorta, including lumen, wall and periaortic region. Additionally, CT scan is more or less exact in the size measurement, and it has a shorter examination time. Three dimensional reconstruction of CT images has an important role in the planning of surgery. However, a CT scan with axial images cannot properly evaluate the very proximal portion of the ascending aorta. In addition, motion artifact can adversely affect the resolution of CT images of the aorta, although technology is improving, especially with ECG gated tomographic angiography. Furthermore, risk of renal damage from the contrast media used during CT scan is a real obstacle in some patients.

Angiography

The shape of the aorta is ideally seen angiographically. Images of the aortic contour are exceptional and morphology of the aortic can be seen beautifully. This can facilitate accurate surgical planning. Additionally, it allow for evaluation and treatment of coronary artery disease, aortic branch disease, as well as assessment of aortic valve and left ventricular function.[97] However, diameter of the ascending aorta from the angiographic images is not always accurate and simple to calculate. It is not available universally because it requires the presence of experienced physician to perform. It has disadvantage of being invasive procedure that is time consuming and require contrast medium with exposure to radiation.

Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) has been recommended as the technique of first choice for the detection and follow-up of both aortic complications and premorbid conditions, such as intramural hematoma, and aortic aneurysm. Its other advantages are ability to assess branch artery involvement, diagnosing aortic valve pathology, and left ventricular dysfunction. MRI is inherently a multiplane modality that can provide high quality images of the aorta in transverse, axial, sagittal, and coronal plane, as well as in left anterior oblique view. MRI has shown high sensibility and specificity both for initial diagnosis and progression of aneurismal disease. MRI offers a non-invasive and accurate evaluation of TAA and it doesn't require nephro-toxic contrast agent or ionizing radiation, although it takes longer time to acquire images and might need sedation in some patients and is not widely available in an emergency basis.[39,42] It is also contraindicated in claustrophobic patients, and patients with metallic prosthesis and pacemakers. Use of cine-MRI techniques, combined to non-invasive haemodynamic data, offers both a morphological and functional examination of the entire aorta, which can provide with information about diameter, geometry, blood flow and aortic wall mechanical properties.[44]

Functional MRI Examination

Most of the study to evaluate ascending aorta using functional MRI has been done in Marfan patients or patients with bicuspid aortic valve.[35] However, its role in evaluating biomechanical property of the ascending aorta in other patients could be valuable as well. Information supplied by functional MRI may have utility in thoracic aortic disease management. Important parameters evaluated by functional MRI are:

Aortic distensibility

The ratio of aortic diameter (or area or volume) and pulse pressure yields a derivative for *in vivo* measurable stiffness, called aortic distensibility.[34] Aortic distensibility can be calculated using functional MRI. To calculate it following formula is used.

$D = (A\,max - A\,min)/[A\,min \times (P\,max\ P\,min)]$

D=distensibility ($10^{-3}mmHg^{-1}$), A max=maximal (systolic) aortic area (mm^2), A min=minimal (diastolic) aortic area (mm^2), P max=systolic blood pressure (mmHg), P min=diastolic blood pressure (mmHg).

Previous studies have shown significantly decrease distensibility of non dilated ascending aorta in Marfan syndrome and bicuspid aortic valve patients when compared to healthy subjects[34,35]. In addition, increasing size of ascending aorta has inverse relationship in determining the distensibility of the aorta.

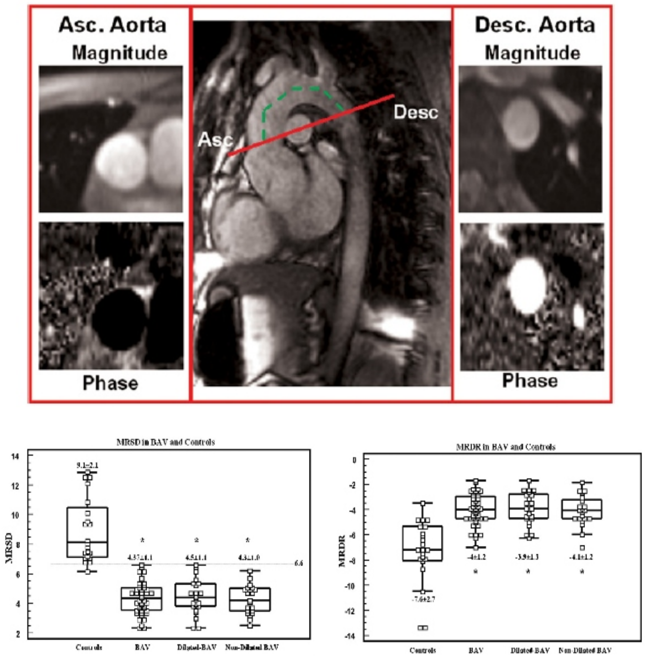
Four dimensional flow study

Given that flow phenomena are important in the hemodynamic etiology of various vascular pathologies, the presence of these flow abnormalities, e.g. blood acceleration, and degradation of measurement accuracy is important to evaluate using functional MRI. The concurrent presentation of 4D velocities and acceleration data demonstrates the potential of this technique to provide a comprehensive measurement of the proximal cardiovascular hemodynamic environment.

Maximal rate of Systolic distension and diastolic recoil

Recently, Aquaro et al has evaluate elastic properties of ascending aorta in young patients under 25 years of age with bicuspid aortic valve using MRI and proposed two new functional MRI indexes, maximal rate of systolic distension (MRSD) and maximal rate of diastolic recoiling (MRDR). As suggested by authors, MRSD is an expression of vessel distension during systole whereas MRDR represents diastolic recoil of the aorta. In these patients with bicuspid aortic valve, a significantly slow MRSD and MRDR has been observed suggesting impaired elastic property of the aorta independent of aortic diameter (Figure 5).

Figure 5: MRSD and MRDR in BAV patients compared to control patient. (Aquaro et al.)



TREATMENT

Gold standard for treatment of the ascending aortic aneurysm is the surgical treatment. However, conservative treatment could be advised at the initial stage with smaller aortic diameter and in patients with morbidity, high risk factors for negative surgical outcome and who are not suitable for surgical treatment due to other coexisting disease.

In adult patients, conservative managements consist of smoking cessation, life style modification, a stringent control of hypertension, lipid profile optimization, and other measure to reduce risk of atherosclerotic process. Furthermore, several other medical treatment options have been studied. Shores et al. in a randomized control study showed that Marfan patients treated daily with Propranolol has slowed aortic root dilation (0.023 vs 0.084 per year). In a recent randomized trial, angiotensin receptor blocker, losartan, added to beta blocker in a group of pediatric population with Marfan syndrome shows more effective protection to slow the progression of aortic root dilatation. Additionally, a matrix metalloproteinase inhibitor, antibiotic doxycycline has shown promising effect in the slowing down of the growth rate of the abdominal aortic aneurysm, but no study has reported effect of doxycycline on ascending aortic aneurysm. To summarize it, none of the studies has shown proven clinical benefit of these medical therapy in the treatment of ascending aortic aneurysm.

Indications For Surgery

If medical therapy can have a palliative role only in high-risk cases, TAA surgery significantly improves 2-year survival from 24% in un-operated patients to 70% in patients undergoing aortic replacement surgery.

All together indications for TAA surgery can be divided into two broad categories.[9]

1. Mandatory indication it includes emergency situations as acute dissection, rupture and intramural haematoma and entails immediate treatment at the time of diagnosis.

2. Elective indication this is basically prophylactic in nature, it is reserved to chronic aneurysm and aims to avoid occurrence of acute, devastating complications.

In the light of currently available surgical techniques, myocardial protection strategies and anesthesia management, elective TAA surgery shows low operative mortality with significant improvement of prognosis at long-term follow-up. Recent series reported an operative mortality of 3-4% for elective operations. This progressive improvement in clinical outcome, during the last two decades, has led surgical community to a revision of the "classic" timing for elective surgery, suggesting more liberal and early indication.

In modern surgical practice, indication for elective TAA surgery depends on two instrumental parameters:

- Absolute diameter of aorta at the time of evaluation.
  Yearly growth rate of aortic diameter, independently from absolute diameter.

1. Absolute Aortic diameter

Absolute aortic size is a key determinant of diagnostic classification and therapeutic management of patients with thoracic aortic aneurysms. Clinical and instrumental criteria to indicate TAA surgery in daily practice actually vary among different surgical centers, according to their experience and operative results. A temporal evolution, towards more aggressive and individualized treatment, has been observed during the last 10-20 years. Elefteriades et al advocated "prophylactic" surgery at the diameter of 55mm for ascending aorta and 65mm for descending aorta aneurysms.[9] This cut-off diameters still represent the most used referral indications for surgery in nowadays practice. Later studies tried to relate surgical timing to patient's characteristics that are known to modify the referral range for "normal diameter" (as body surface area, sex and age) or influence the risk of sudden complications (aneurysm etiology, familial history of rupture or dissection, concomitant valve incompetence, etc). Ergin and co-workers suggest the ratio between measured diameter/predicted diameter (according to sex, age and body surface area) to be more effective than absolute size evaluation for surgical timing.[9] Concomitant risk factors for rupture or dissection (Marfan syndrome, bicuspid aortic valve, severe aortic regurgitation, aneurysm-related death in familial history, female sex, etc) usually lead to an earlier indication to surgery, with aortic ratio between 1.3 to 1.4.[9] Body mass index also affected aortic diameter by 0.27 mm (0.14 to 0.44 mm) per unit of body mass index. Taking into consideration of these evident, in 2006, the group from Yale University proposed a new parameter for better stratifying TAA patients according to the risk of rupture, dissection or aneurysm related death: an "Aortic Size Index (ASI)" (aortic absolute diameter / body surface area) of 2.75 cm/ m^2 or more should prompt to elective TAA surgery to avoid moderate to severe (8-20%) yearly risk of adverse events (Table 1).[103] By proportional hazard analysis of rupture, they have shown that the risk of rupture is 11 times higher with aortic size index 4.25 to 4.99 cm/m^2 than with ASI 2.00 to 2.74 cm/m^2. This means that, in contrast to "classic" indications, smaller patients may benefit from operative repair at even smaller sizes. For example, a 45mm aneurysm in a small lady, with body surface area (BSA) of 1.4m^2, should be nowadays surgically treated.[96]

Table 1: Aortic Size Index chart (Davies et al.)

BSA	Aortic size (cm) 3.5	4.0	4.5	5.0	5.5	6.0	6.5	7.0	7.5	8.0
1.30	2.69	3.08	3.46	3.85	4.23	4.62	5.00	5.38	5.77	6.15
1.40	2.50	2.86	3.21	3.57	3.93	4.29	4.64	5.00	5.36	5.71
1.50	2.33	2.67	3.00	3.33	3.67	4.00	4.33	4.67	5.00	5.33
1.60	2.19	2.50	2.80	3.13	3.44	3.75	4.06	4.38	4.69	5.00
1.70	2.05	2.35	2.65	2.94	3.24	3.53	3.82	4.12	4.41	4.71
1.80	1.94	2.22	2.50	2.78	3.06	3.33	3.61	3.89	4.17	4.44
1.90	1.84	2.11	2.37	2.63	2.89	3.16	3.42	3.68	3.95	4.22
2.00	1.75	2.00	2.25	2.50	2.75	3.00	3.25	3.50	3.75	4.00
2.10	1.67	1.90	2.14	2.38	2.62	2.86	3.10	3.33	3.57	3.80
2.20	1.59	1.82	2.05	2.27	2.50	2.72	2.95	3.18	3.41	2.64
2.30	1.52	1.74	1.96	2.17	2.39	2.61	2.83	3.04	3.26	3.48
2.40	1.46	1.67	1.88	2.08	2.29	2.50	2.71	2.92	3.13	3.33
2.50	1.40	1.60	1.80	2.00	2.20	2.40	2.60	2.80	3.00	3.20

= low risk (~4% per yr)
= moderate risk (~8% per yr)
= severe risk (~20% per yr)

2. Aortic growth rate

Normal aorta expands about 1-2 mm over 10 years. The expansion rate of the aneurysm in the ascending aorta is about

1.3 mm/year, with a greater rate of 3.9 mm/year for descending aorta. The growth rate is proportional to aneurysm diameter. One study has demonstrated that in patients with ascending aorta aneurysms, with initial diameter 35-40 mm show a mean yearly progression of 2.1mm, in contrast to 5.6 mm yearly rate for aneurysm larger than 60mm Considering that rapid progression of aneurysm size as a marker of ominous events, elective surgery is generally indicated if dilation rate of pathological aorta is > 10mm per year, independently from absolute aortic diameter.

Surgical procedure for the treatment of Ascending aortic aneurysms

Surgical treatment of the ascending aortic aneurysm compromises endovascular grafting and open surgical procedure. The decision to treat an aneurysm must be made with same rigor for endovascular therapy as for open surgical therapy. The presence of a small thoracic aneurysm is not a valid indication for endovascular therapy just because stent therapy is available. Furthermore, stent deployment for the ascending aortic aneurysm is not approved in most of the countries.

Recommendation for open surgery for ascending aortic aneurysm

Asymptomatic patients with degenerative thoracic aneurysm who are otherwise suitable candidates and for whom the ascending aorta or aortic sinus diameter is 5.5 cm or greater, should be evaluated for surgical repair.

Patients with Marfan syndrome or other genetically mediated disorders (Ehler-Danlos syndrome, Turner syndrome, bicuspid aortic valve) should undergo elective surgery at smaller diameter, i.e. 4.0 to 5.0 cm depending on the condition, to avoid acute dissection or rupture.

Surgical Procedures

Separate valve and ascending aortic replacement are recommended in patients without significant aortic root dilatation, in elderly patients, or in young patients with minimal dilatation who have aortic valve disease. (*Level of Evidence: C*)

Patients with Marfan, Loeys-Dietz, and Ehlers-Danlos syndromes and other patients with dilatation of the aortic root and sinuses of Valsalva should undergo excision of the sinuses in combination with a modified David reimplantation operation if technically feasible or, if not, root replacement with valved graft conduit. (*Level of evidence: B*). The extent of ascending aortic resection are determined by preoperative imaging and intraoperative findings. For patients with isolated ascending aortic aneurysm, resection and Dacron graft replacement is the recommended procedure.

Whereas, for patients with aortic stenosis requiring valve replacement, the choice of valve is determined by age of the patient, life expectancy, risk of complications related to anticoagulation and reoperation. For patients with aortic regurgitation, repair of the aortic valve with or without root remodeling is preferable if the valve is not severely fibrotic or calcified.

In patients with a dilated aortic root, particularly those with stenotic bicuspid valves, composite valve graft containing either mechanical or biological aortic prosthesis are implanted (Bentall Procedure). However, in elderly patients, sometime ascending aortic aortoplasty may be an acceptable option, if the aortic diameter doesn't exceed 5.0 cm.

In patients with aortic regurgitation and ascending aortic aneurysm, aortic valve repair with root-sparing procedure may be the primary option to consider (Tiron Davide operation). Similarly, Marfan patients with tricuspid aortic valve regurgitation might benefit from a modification of David reimplantation surgery.

CONCLUSION

Ascending aortic aneurysm is a highly lethal and indolent disease. Usually aneurysmatic aorta grows asymptomatically until it dissects or rupture. Pathophysiology of ascending aortic aneurysm is a complex process. Diagnosis of the ascending aortic aneurysm is made incidentally. So, a high degree of suspicion and diagnostic screening using non-invasive imaging methods allow to detect and treat these vulnerable patients earlier. Conservative treatment could be advised at the initial stage with smaller aortic diameter and in patients with morbidity, high risk factors for negative surgical outcome and who are not suitable for surgical treatment due to other coexisting disease.

Surgical treatment is the gold standard for symptomatic patients and asymptomatic patients with aortic diameter more than 5.5 cm in otherwise normal patient, while 4.0-5.0 cm in Marfan patients and patients with genetically mediated process. Postoperative morbidity and mortality has significantly decreased due to better anesthetic management, improved surgical techniques and progress in preoperative and postoperative care.

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