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DISEASES OF THE AORTA

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2-я КАФЕДРА ХИРУРГИЧЕСКИХ БОЛЕЗНЕЙ

А. В. Жура, В. Я. Хрыщанович

ЗАБОЛЕВАНИЯ АОРТЫ

DISEASES OF THE AORTA

Учебно-методическое пособие

Минск БГМУ 2016

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Приведены сведения о наиболее часто встречающихся заболеваний аорты. Подробно изложены методы диагностики заболеваний аорты, вопросы этиологии и патогенеза в соответствии с международными классификациями, основные осложнения и методы их коррекции, современные направления хирургического лечения.

Предназначено для студентов 4–6-го курсов медицинского факультета иностранных учащихся, обучающихся на английском языке.

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MOTIVATIONAL CHARACTERISTIC OF THE TOPIC

Total in-class hours: 2.

Diseases of the aorta account for significant cardiovascular morbidity and mortality worldwide. The incidence of aortic diseases is expected to rise with the increasing age of the population. Diagnostic evaluation of aortic disorders has improved in the last 2 decades, allowing earlier diagnosis and therapeutic intervention. This issue summarizes the major disease entities affecting the aorta, and reviews the current practice guidelines on their evaluation and management.

The purpose is to study the main surgical diseases of aorta, etiology, clinical presentations, diagnostics and principles of treatment.

Objectives are:

1. To learn current methods of investigations in aortic diseases.
2. To learn main etiology, clinical picture, diagnosis and treatment of aortic aneurysm.
3. To learn main etiology, clinical picture, diagnosis of aortic dissection.
4. To know the indications for surgery for acute and chronic aortic diseases.
5. To know the principles and steps of surgical repair for acute and chronic aortic diseases.

Requirements for the initial knowledge level.

To learn the topic completely the student must know:

- propaedeutics of internal diseases (methods of clinical evaluations of abdominal organs);
- human anatomy (embryogenesis, localization and structure of a cardiovascular system);
- topographic anatomy and operative surgery (main surgical approaches to thoracic and abdominal aorta);
- normal physiology (functions of cardiovascular system);
- general surgery (basic principles of hemorrhage).

Test questions from related disciplines:

1. Normal and topographic anatomy of aorta.
2. Embryogenesis of main arterial vessels.
3. Function of a cardiovascular system.
4. Clinical evaluation of cardiovascular system pathology.
5. Surgical approaches to aorta and heart.

Test questions:

1. Anatomical data.
2. Diagnostic studies.
3. Thoracic aortic aneurysm. Etiology. Pathogenesis. Classification. Clinical picture. Treatment.
4. Dissection of the aorta. Etiology. Pathogenesis. Classification. Clinical picture. Treatment.
5. Abdominal aortic aneurysm. Etiology. Pathogenesis. Classification. Clinical picture. Treatment.

STUDY MATERIAL

ANATOMY

The aorta is the main conduit and reservoir of oxygenated blood in the body. A large elastic artery, it is composed of three layers: The intima is the innermost layer and includes the single-layered endothelium. The media is the thickest layer of the aortic wall and is composed of sheets of elastic tissue, smooth muscle cells, and collagen, which provide the aorta with its tensile strength and distensibility. The adventitia is the outermost layer; it is composed of loose connective tissue and contains the vasa vasorum, which constitutes the blood supply to the aortic wall.

The aorta consists of two major segments — the *thoracic aorta* and the *abdominal aorta* (fig. 1). The thoracic aorta consists of the aortic root, from the aortic annulus, including the sinuses of Valsalva, up to the level just above the sinotubular junction; the ascending aorta, from the sinotubular junction to the innominate artery, with an average diameter of 3 cm; the arch, from the innominate artery to the left subclavian artery; and the descending thoracic aorta, with an average diameter of 2.5 cm, which begins after the origin of the left subclavian artery. The abdominal aorta begins when the descending thoracic aorta passes through the diaphragm. The abdominal aorta (average diameter, 2.0 cm) is further classified as suprarenal or infrarenal according to origin of renal arteries.

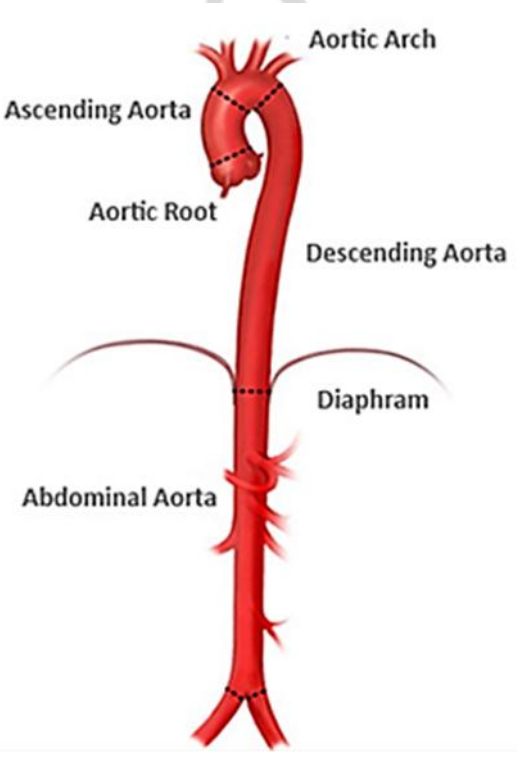
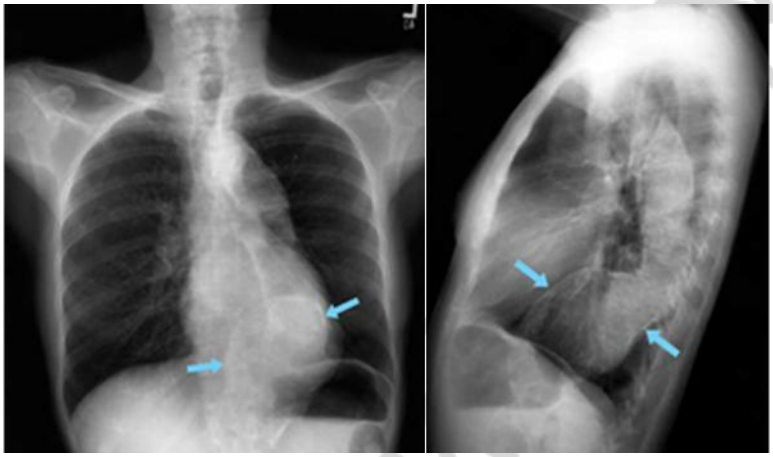


Fig 1. Segments of the aorta

METHODS OF INVESTIGATION

PLAIN RADIOGRAPHY

X-Ray film made in the direct or lateral projection may show aortic aneurysms as a convex shadow to the right or to the left of the cardiac silhouette (fig. 2). Aortic calcification also may be seen in the upper abdomen on a standard radiograph. But it is important to recognize that chest radiographs often appear normal in patients with thoracic aortic disease and thus cannot exclude the diagnosis of aortic aneurysm.



a ***b***

Fig. 2. Chest x-ray of a patient with a thoracic aortic aneurysm in direct (*a*) and lateral (*b*) positions

Abdominal aortic aneurysms are often discovered on abdominal or pelvic scans done for other indications (e. g., back pain or renal cysts). Plain films of the lumbar region may show a calcified shell of the aorta.

ECHOCARDIOGRAPHY

Both transthoracic and transesophageal echocardiography provide excellent visualization of the ascending aorta, including the aortic root. Transesophageal echocardiography also allows visualization of the descending thoracic aorta but is not ideal for evaluating the transverse aortic arch (which is obscured by air in the tracheobronchial tree) or the upper abdominal aorta. Effective echocardiography requires considerable technical skill, both in obtaining adequate images and in interpreting them.

During echocardiography the ascending aortic aneurysms are commonly discovered in patients presenting with symptoms or signs of aortic valve regurgitation. It is possible to discover the aortic dissection.

ULTRASOUND

Real-time (***grey-scale***) or B-mode ultrasonography (***duplex ultrasound***) is a non-invasive investigation that gives anatomical detail of the aortic wall, velocity and direction of blood flow, presence of thrombi etc. It also provides an accurate measurement of aneurysm size and revealing of hematoma around ruptured aneurysm.

COMPUTED TOMOGRAPHY/MAGNETIC RESONANCE ANGIOGRAPHY

Computed tomographic (CT) scanning provides visualization of the entire thoracic and abdominal aorta. Consequently, CT is the most common—and arguably the most useful—imaging modality for evaluating aortic aneurysms. Systems capable of constructing multiplanar images and performing three-dimensional aortic reconstructions (fig. 3) are widely available. In addition to establishing the diagnosis, CT provides information about an aneurysm's location, extent, anatomic anomalies, and relationship to major branch vessels. CT is particularly useful in determining the absolute diameter of the aorta, especially in the presence of laminated clot. Contrast-enhanced CT (CTA, computed tomographic angiography) as a magnetic resonance angiography (MRA) provides information about the aortic lumen and can detect mural thrombus, aortic dissection, inflammatory periaortic fibrosis, and mediastinal or retroperitoneal hematoma due to contained aortic rupture.

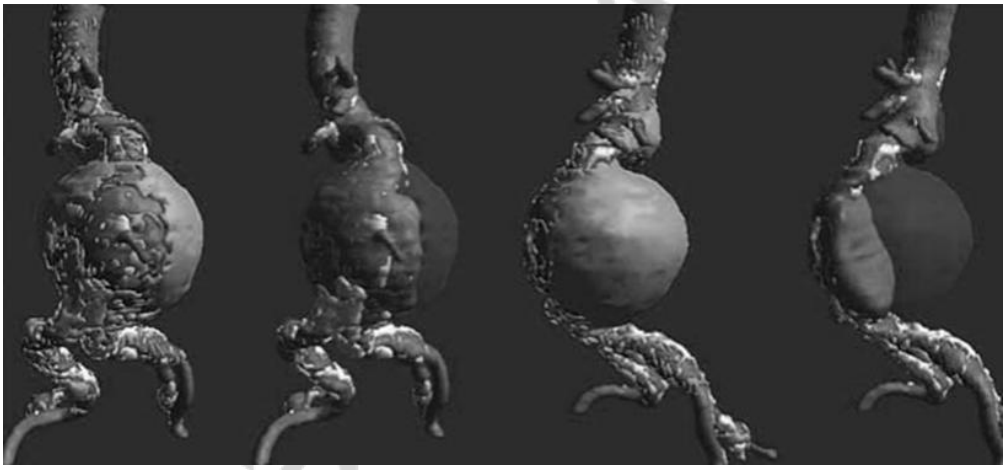
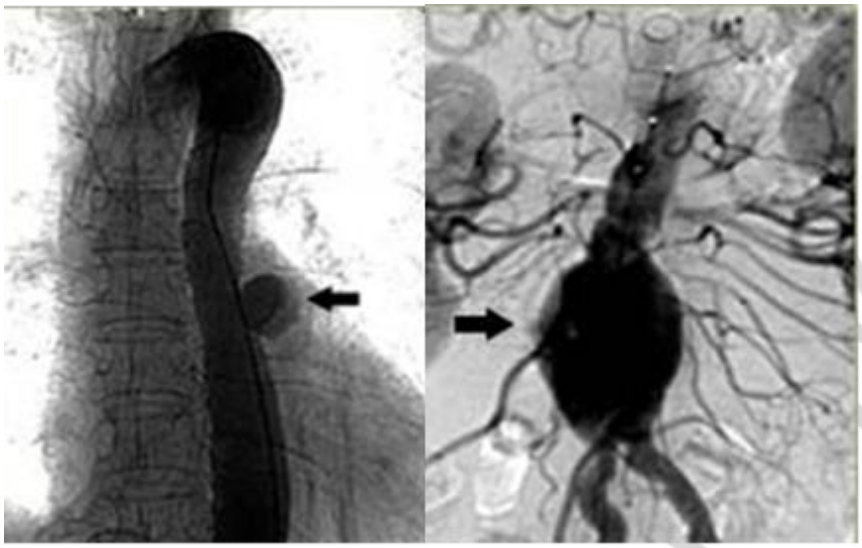


Fig. 3. Three dimensional reconstructions of abdominal aorta aneurysm during CT angiography

ANGIOGRAPHY

Although diagnostic aortography (fig. 4) was, until recently, considered the gold standard for evaluating aortic diseases, CTA and MRA have largely replaced this modality. Technologic improvements have enabled CTA and MRA to provide excellent aortic imaging while causing less morbidity than catheter-based studies do, so *CTA and MRA should now be considered the gold standard*. Therefore, the role of diagnostic angiography in patients with aortic diseases is currently limited.

However, the advent of endovascular therapies has given catheter-based angiography a new role, because intraprocedural angiography is an essential component of endovascular procedures.



a ***b***

Fig. 4. Angiography of aorta; *a* — saccular descending aortic aneurysm; *b* — abdominal aortic aneurysm

THORACIC AORTIC ANEURYSMS

Aortic aneurysm is defined as a permanent, localized or diffused dilatation of the aorta to a diameter that is at least 50% greater than is normal at that anatomic level.

ETIOLOGY AND PATHOGENESIS

The normal aorta derives its elasticity and tensile strength from the medial layer, which contains approximately 45 to 55 lamellae of elastin, collagen, smooth muscle cells, and ground substance. Elastin content is highest within the ascending aorta, as would be expected because of its compliant nature, and decreases distally into the descending and abdominal aorta. Maintenance of the aortic matrix involves complex interactions among smooth muscle cells, macrophages, proteases, and protease inhibitors. Any alteration in this delicate balance can lead to aortic disease.

Thoracic aortic aneurysms have a variety of causes (see the list below). Although these disparate pathologic processes differ in biochemical and histologic terms, they share the final common pathway of progressive aortic expansion and eventual rupture.

Causes of thoracic aortic aneurysms:

1. Nonspecific medial degeneration.

It is the most common cause of thoracic aortic disease. Histologic findings of mild medial degeneration, including fragmentation of elastic fibers and loss of smooth muscle cells, are expected in the aging aorta. However, an advanced, accelerated form of medial degeneration leads to progressive weakening of the aortic wall, aneurysm formation, and eventual dissection, rupture, or both. The underlying causes of medial degenerative disease remain unknown.

2. **Aortic dissection (see chapter Aortic Dissection).**
3. **Genetic disorders.**

Marfan syndrome (fig. 5). Marfan syndrome is an autosomal dominant genetic disorder characterized by a specific connective tissue defect that leads to aneurysm formation. The phenotype of patients with Marfan syndrome typically includes a tall stature, high palate, joint hypermobility, eye lens disorders, mitral valve prolapse, and aortic aneurysms. Between 75 and 85 % of patients with Marfan syndrome have dilatation of the ascending aorta and annuloaortic ectasia (dilatation of the aortic sinuses and annulus).

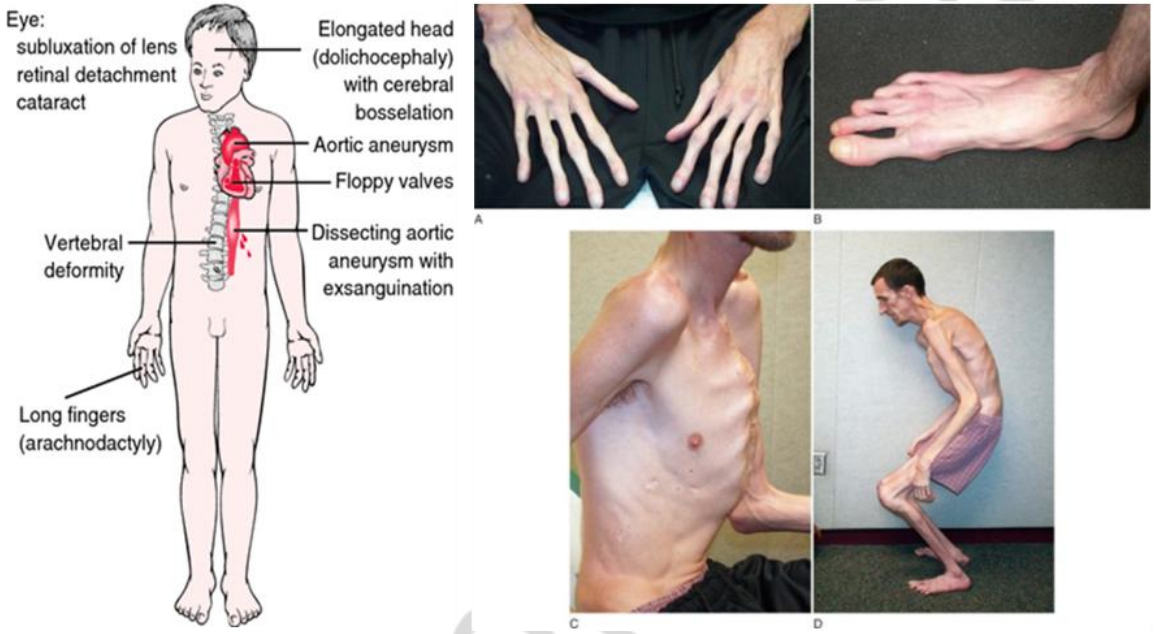


Fig. 5. Clinical manifestations of Marfan syndrome

Loeys-Dietz syndrome. It is characterized as an aneurismal syndrome with widespread systemic involvement. Loeys-Dietz syndrome is an aggressive, autosomal dominant condition that is distinguished by the triad of arterial tortuosity and aneurysms, hypertelorism (widely spaced eyes), and bifid uvula or cleft palate.

Ehlers-Danlos syndrome. Ehlers-Danlos syndrome includes a spectrum of inherited connective tissue disorders of collagen synthesis.

Familial aortic aneurysms. Families without the heritable connective tissue disorders. In fact, it is estimated that at least 20% of patients with thoracic aortic aneurysms and dissections have a genetic predisposition to them.

Congenital bicuspid aortic valve. Bicuspid aortic valve is the most common congenital malformation of the heart or great vessels. patients with bicuspid aortic valves have an increased incidence of ascending aortic aneurysm formation and, often, a more rapid rate of aortic enlargement. This dilatation usually is limited to the ascending aorta and root.

4. **Poststenotic dilatation.**
5. **Infection.**

In other parts of the world syphilitic aneurysms remain a major cause of morbidity and mortality. The spirochete *Treponema pallidum* causes an obliterative endarteritis of the vasa vasorum that results in medial ischemia and loss of the elastic and muscular elements of the aortic wall. The ascending aorta and arch are the most commonly involved areas. The mycotic infection also may induce the formation of thoracic aortic aneurysm.

6. Aortitis

Systemic autoimmune disorders may cause thoracic aortitis:

- Takayasu's arteritis
- Giant cell arteritis
- Rheumatoid aortitis

7. Trauma

Traumatic pseudoaneurysms of the thoracic aorta usually represent chronic leaks that are contained by surrounding tissue and fibrosis. By definition, the wall of a pseudoaneurysm is not formed by intact aortic tissue; rather, the wall develops from organized thrombus and associated fibrosis.

CLASSIFICATION

According to involvement of aortic segments, aneurysms of the thoracic aorta are divided to:

- Root aneurysms;
- Aneurysm of the ascending aorta;
- Aneurysm of the aortic arch;
- Descending thoracic aortic aneurysms;
- Thoracoabdominal aneurysms, that may be of 4 types (fig. 6).

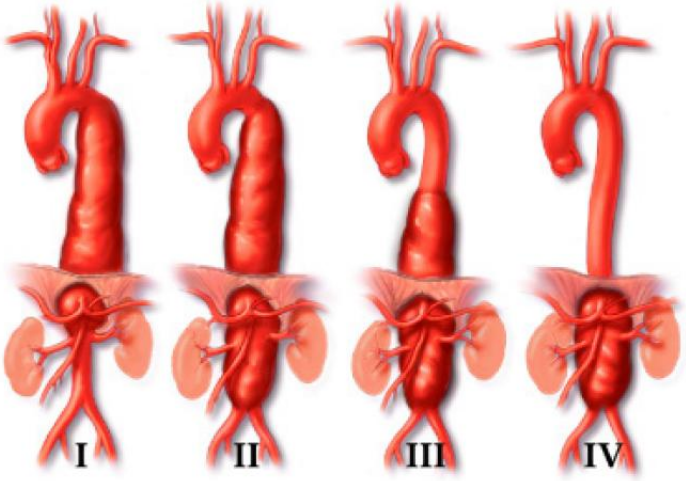


Fig. 6. Crawford classification of thoracoabdominal aortic aneurysms

According to structure, aortic aneurysms can be **true** or **false.**

True aneurysms can take two forms: fusiform and saccular (fig. 7). Fusiform aneurysms are more common and can be described as symmetrical dilatations of the aorta. Saccular aneurysms are localized outpunching of the aorta.

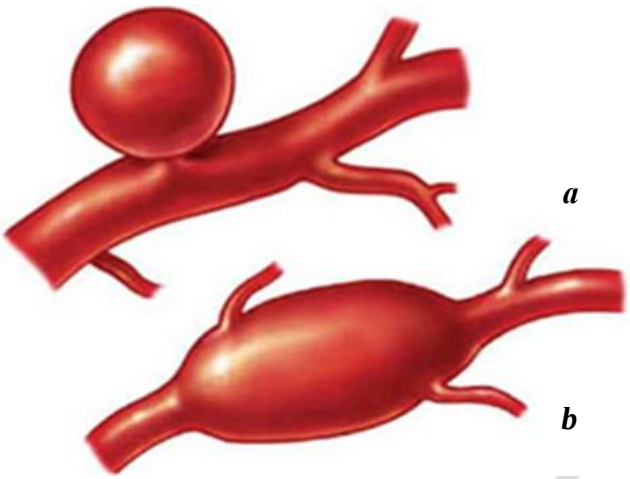


Fig. 7. Aortic aneurysm by shape. *a* — saccular, *b* — fusiform

False aneurysms, also called pseudoaneurysms, are leaks in the aortic wall that cause blood to collect in pouches of scar tissue on the exterior of the aorta (pulsatile hematoma).

NATURAL HISTORY

Clinical course of thoracic aortic aneurysm classically is characterized as ***progressive aortic dilatation*** and eventual ***dissection***, ***rupture***, or both.

Aortic diameter was a strong predictor of rupture, dissection, and mortality. Critical diameters, at which the incidence of expected complications significantly increased, were 6.0 cm for aneurysms of the ascending aorta and 7.0 cm for aneurysms of the descending thoracic aorta; the corresponding risks of rupture after reaching these diameters were 31 % and 43 % respectively.

Certain types of aneurysms have an increased propensity for expansion and rupture. For example, aneurysms in patients with Marfan syndrome dilate at an accelerated rate and rupture or dissect at smaller diameters than non–Marfan-related aneurysms. Before the era of surgical treatment for aortic aneurysms, this aggressive form of aortic disease resulted in an average life expectancy of 32 years for Marfan patients.

CLINICAL MANIFESTATIONS

In many patients with thoracic aortic aneurysms, the aneurysm is discovered incidentally when imaging studies are performed for unrelated reasons. Therefore, patients often are asymptomatic at the time of diagnosis. However, thoracic aortic aneurysms that initially go undetected eventually create symptoms and signs that correspond with the segment of aorta that is involved. These aneurysms have a wide variety of manifestations, including compression or erosion of adjacent structures, aortic valve regurgitation, distal embolism, and rupture.

Local compression and erosion syndrome .Initially, aneurysmal expansion and impingement on adjacent structures, causes mild chronic pain. The most common symptom in patients with ascending aortic aneurysms is anterior chest

discomfort; the pain is frequently precordial in location but may radiate to the neck and jaw, mimicking angina. Aneurysms of the ascending aorta and transverse aortic arch can cause symptoms related to compression of the superior vena cava, the pulmonary artery, the airway, or the sternum. Rarely, these aneurysms erode into the superior vena cava or right atrium, causing acute high-output failure. Expansion of the distal aortic arch can stretch the recurrent laryngeal nerve, which results in left vocal cord paralysis and hoarseness. Descending thoracic and thoracoabdominal aneurysms frequently cause back pain localized between the scapulae. When the aneurysm is largest in the region of the aortic hiatus, it may cause middle back and epigastric pain.

Descending thoracic aortic aneurysms may cause varying degrees of airway obstruction, manifesting as cough, wheezing, stridor, or pneumonitis. Pulmonary or airway erosion presents as hemoptysis. Compression and erosion of the esophagus cause dysphagia and hematemesis, respectively.

Aortic valve regurgitation. Ascending aortic aneurysms can cause displacement of the aortic valve commissures and annular dilatation. The resulting deformation of the aortic valve leads to progressively worsening aortic valve regurgitation. In response to the volume overload, the heart remodels and becomes increasingly dilated. Patients with this condition may present with progressive heart failure, a widened pulse pressure, and a diastolic murmur.

Distal embolization. Thoracic aortic aneurysms — particularly those involving the descending and thoracoabdominal aorta — are commonly lined with friable, atheromatous plaque and mural thrombus. This debris may embolize distally, causing occlusion and thrombosis of the visceral, renal, or lower-extremity branches.

Rupture. Patients with ruptured thoracic aortic aneurysms often experience sudden, severe pain in the anterior chest (ascending aorta), upper back or left chest (descending thoracic aorta), or left flank or abdomen (thoracoabdominal aorta). When ascending aortic aneurysms rupture, they usually bleed into the pericardial space, producing acute cardiac tamponade and death. Descending thoracic aortic aneurysms rupture into the pleural cavity, producing a combination of severe hemorrhagic shock and respiratory compromise. External rupture is extremely rare; saccular syphilitic aneurysms have been observed to rupture externally after eroding through the sternum.

TREATMENT

Thoracic aortic aneurysms should be repaired to prevent fatal rupture. Prophylactic operation is recommended when the diameter of an ascending aortic aneurysm is >5.5 cm, when the diameter of a descending thoracic aortic aneurysm is >6.5 cm, or when the rate of dilatation is >1 cm/year. In patients with connective tissue disorders, such as Marfan and Loeys-Dietz syndromes, the threshold for operation is lower with regard to both absolute size (5.0 cm for the ascending aorta and 6.0 cm for the descending thoracic aorta) and rate of growth.

Smaller ascending aortic aneurysms (4.0 to 5.5 cm) also are considered for repair when they are associated with significant aortic valve regurgitation.

Open repair. Operations to repair aortic aneurysms require cardiopulmonary bypass. The best choice of aortic replacement technique varies depending on the extent of the aneurysm and the condition of the aortic valve. The spectrum of operations ranges from simple graft replacement of the tubular portion of the ascending aorta to graft replacement of the entire proximal aorta, including the aortic root, and reattachment of the coronary arteries and brachiocephalic branches (fig. 8).

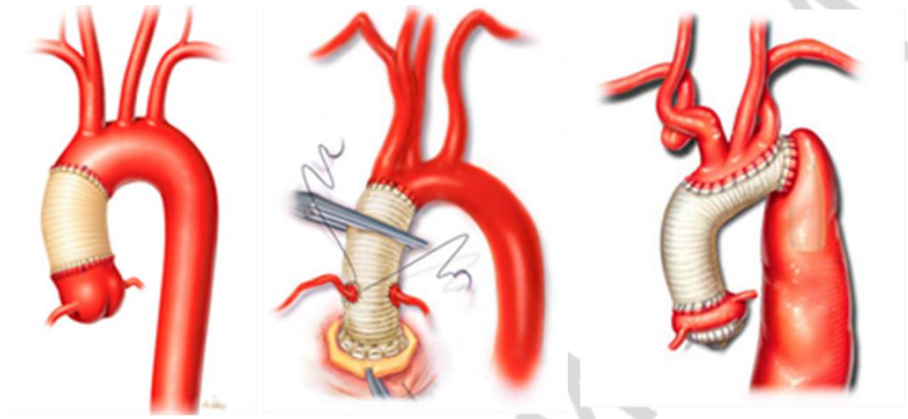


Fig. 8. Variants of open repair of thoracic aneurysms

Endovascular Repair. Endovascular repair of thoracic aortic aneurysms (fig. 9) has become an accepted treatment option in selected patients, particularly patients with isolated degenerative descending thoracic aortic aneurysms. Experience with purely endovascular treatment of proximal aortic disease remains limited and only investigational.

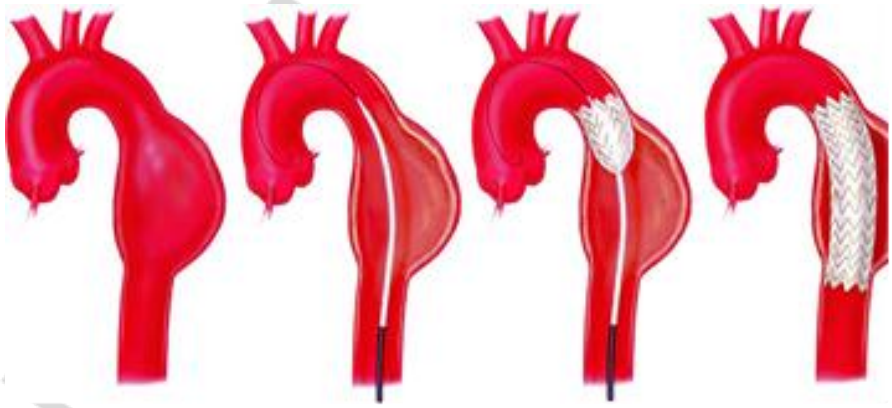


Fig. 9. Steps of endovascular repair

AORTIC DISSECTION

Aortic dissection, the most common catastrophic event involving the aorta, is a progressive separation of the aortic wall layers that usually occurs after a tear forms in the intima and inner media. As the separation of the layers of the media propagates, at least two channels form: the *original lumen*, which remains lined by

the intima and which is called the true lumen, and the newly formed channel within the layers of the media, which is called the *false lumen*. The dissecting membrane separates the true and false lumens (fig. 10). Additional tears in the dissecting membrane that allow communication between the two channels are called re-entry sites. The separation of layers primarily progresses distally along the length of the aorta.

Fig. 10. Dissection of aorta

The extensive disruption of the aortic wall has severe anatomic consequences. *First*, the outer wall of the false lumen is extremely thin, inflamed, and fragile, which makes it prone to ***expansion*** or ***rupture*** in the face of ongoing hemodynamic stress.

Second, the expanding false lumen can compress the true lumen and cause ***malperfusion syndrome*** by interfering with blood flow in the aorta or any of its branch vessels, including the coronary, carotid, intercostal, visceral, renal, and iliac arteries (fig. 11).

Finally, when the separation of layers occurs within the aortic root, the aortic valve commissures can become unhinged, which results in ***acute valvular regurgitation*** (fig. 12).

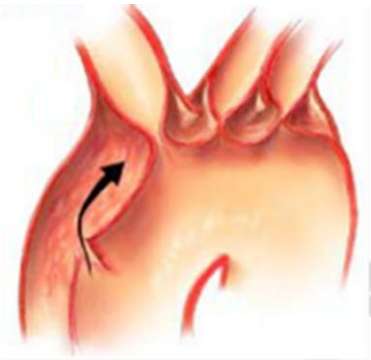


Fig. 11. Compression the origin of brachiocephalic, carotid and subclavian arteries by «false» lumen

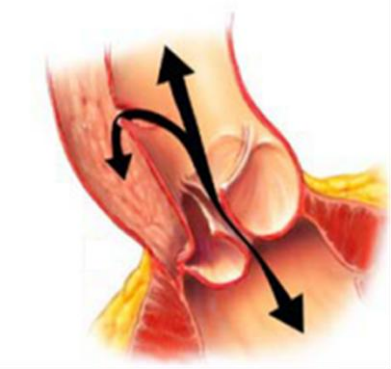


Fig. 12. Acute regurgitation in the aortic valve due to the aortic dissection

Dissection vs. Aneurysm

The relationship between dissection and aneurysmal disease requires clarification. *Dissection and aneurysm are separate entities*, although they often coexist and are mutual risk factors**. *In most cases, dissection occurs in patients without aneurysms***. The subsequent progressive dilatation of the weakened outer aortic wall results in an aneurysm. On the other hand, in patients with degenerative aneurysms, the ongoing deterioration of the aortic wall can lead to a superimposed dissection. The overused term dissecting aneurysm should be reserved for this specific situation.

CLASSIFICATION

Classification for aortic dissection is based on which portions of the aorta are involved (fig. 13). Dissection can be confined to the ascending aorta (left) or descending aorta (middle), or it can involve the entire aorta (right).

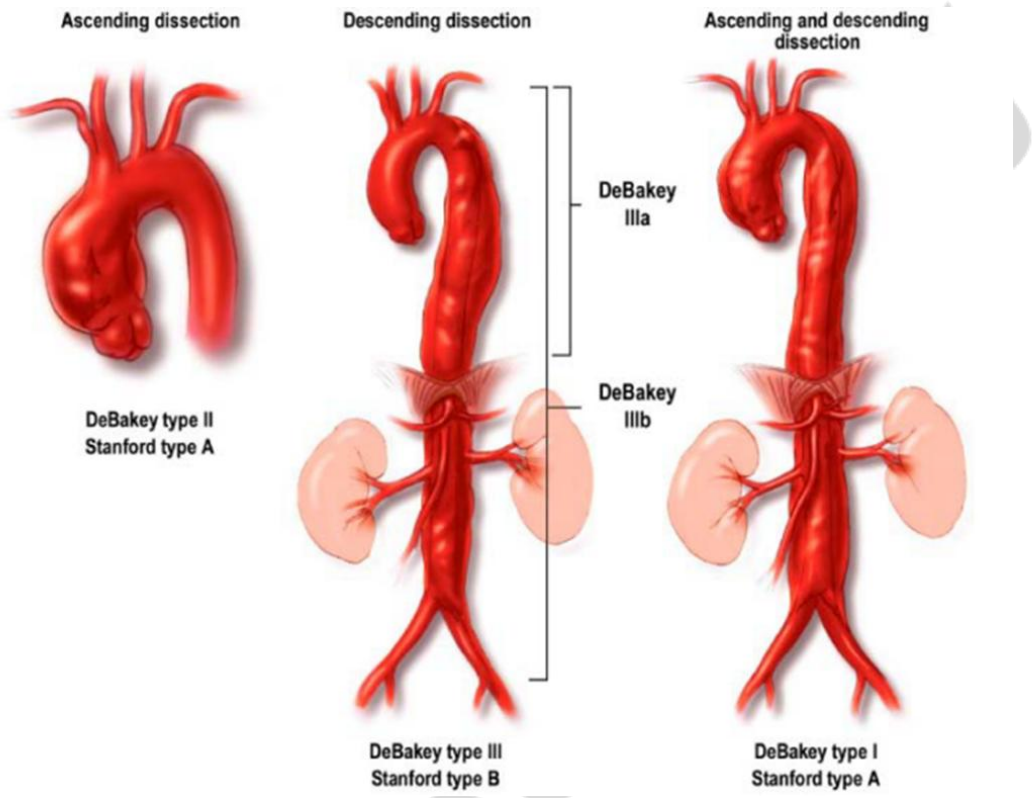


Fig. 13. Stanford and DeBakey classifications schemes of aortic dissection

In addition, aortic dissection is categorized according to the time elapsed since the initial tear. Dissection is considered ***acute*** within the first 14 days after the initial tear; after 14 days, the dissection is considered ***chronic***.

ETIOLOGY AND NATURAL HISTORY

Aortic dissection is a lethal condition with a reported incidence of 3.5 per 100,000. Without appropriate modern medical or surgical treatment, most patients (approximately 90 %) die within 3 months of dissection, mostly from rupture.

Although several risk factors for aortic dissection have been identified, the specific causes remain unknown. Ultimately, any condition that weakens the aortic wall increases the risk of aortic dissection. Common general cardiovascular risk factors, such as smoking, hypertension, atherosclerosis, and hypercholesterolemia, are associated with aortic dissection. Patients with connective tissue disorders, aortitis, bicuspid aortic valve, aortic coarctation, or pre-existing medial degenerative disease are at risk for dissection, especially if they already have a thoracic aortic aneurysm. Aortic injury during cardiac catheterization or surgery is a common cause of iatrogenic dissection. Other situations that are associated with aortic dissection include cocaine abuse and amphetamine abuse.

CLINICAL MANIFESTATIONS

The onset of dissection often is associated with ***severe chest or back pain***, classically described as «tearing», that migrates distally as the dissection progresses along the length of the aorta. The location of the pain often indicates which aortic segments are involved. Pain in the anterior chest suggests involvement of the ascending aorta, whereas pain in the back and abdomen generally indicates involvement of the descending and thoracoabdominal aorta.

Other complications of dissection of the aorta (and involved secondary arteries) are highly varied, including, but not limited to, cardiac ischemia (coronary artery) or tamponade, stroke (brachiocephalic arteries), paraplegia or paraparesis (intercostal arteries), mesenteric ischemia (superior mesenteric artery), kidney failure (renal arteries), and limb ischemia or loss of motor function.

DIAGNOSTIC EVALUATION

Because of the variations in severity and the wide variety of potential clinical manifestations, the diagnosis of acute aortic dissection can be challenging. Diagnostic delays are common; delays beyond 24 hours after hospitalization occur in up to 39% of cases. Unfortunately, delays in diagnosis lead to delays in treatment, which can have disastrous consequences.

Once the diagnosis of dissection is considered, the thoracic aorta should be imaged with contrast-enhanced CT (fig. 14), MRA, or echocardiography.

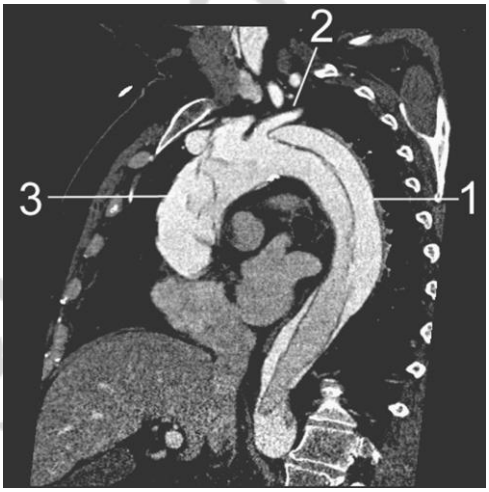


Fig. 14. Computed tomography of aortic dissection; *1* — false lumen; *2* — origin of carotid artery; *3* — dissection in the aortic root

TREATMENT

Because of the potential for rupture before the diagnosis is confirmed, aggressive pharmacologic management is started once there is clinical suspicion of dissection, and this treatment is continued during the diagnostic evaluation. *The goals of pharmacologic treatment are to stabilize the dissection and prevent rup-*

ture. Patients are monitored closely in an intensive care unit. Central venous catheters assure reliable intravenous access for delivering vasoactive medications.

Nonoperative, pharmacologic management of acute descending aortic dissection results in lower morbidity and mortality rates than surgical treatment does. The most common causes of death during nonoperative treatment are aortic rupture and end-organ malperfusion. Therefore, patients are continually reassessed for new complications. At least two serial CT scans—usually obtained on day 2 or 3 and on day 8 or 9 of treatment—are compared with the initial scan to rule out significant aortic expansion. Once the patient's condition has been stabilized, pharmacologic management is gradually shifted from intravenous to oral medications. Oral therapy, usually including a beta antagonist, is initiated when systolic pressure is consistently between 100 and 110 mmHg and the neurologic, renal, and cardiovascular systems are stable. Many patients can be discharged after their blood pressure is well controlled with oral agents and after serial CT scans confirm the absence of aortic expansion.

Long-term pharmacologic therapy is important for patients with chronic aortic dissection. Beta blockers remain the drugs of choice.

Operative treatment. *Acute ascending aortic dissection has traditionally been considered an absolute indication for emergency surgical repair* (fig. 15). However, specific patient groups may benefit from nonoperative management or delayed operation. Delayed repair should be considered for patients who (*a*) present with acute stroke or mesenteric ischemia, (*b*) are elderly and have substantial comorbidity, (*c*) are in stable condition and may benefit from transfer to specialized centers, or (*d*) have undergone a cardiac operation in the remote past.

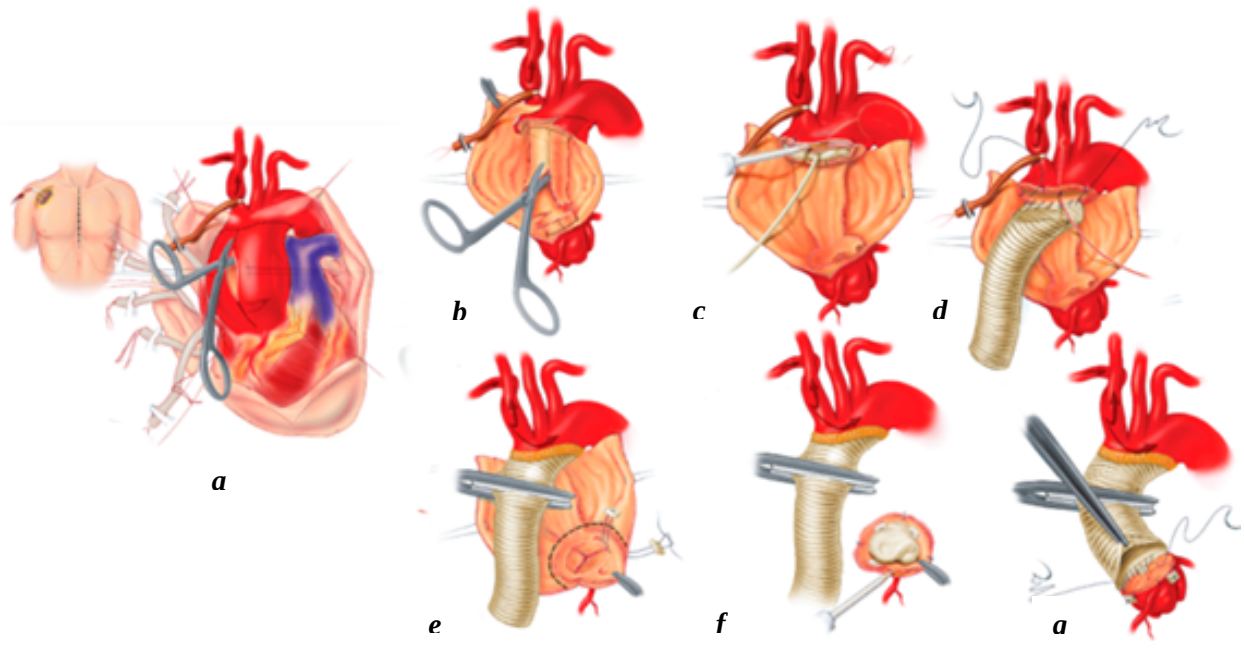


Fig. 15. Steps (A–G) of open repair of the ascending aortic dissection

Occasionally, patients with ascending aortic dissection present for repair in the chronic phase. In most respects, the operation is similar to that for acute dissection repair.

Surgical intervention for *acute descending aortic dissection* is intended to prevent or repair ruptures and relieve ischemic manifestations. During the acute phase of a dissection, *the specific indications for operative intervention include aortic rupture, increasing periaortic or pleural fluid volume, rapidly expanding aortic diameter, uncontrolled hypertension, and persistent pain despite adequate medical therapy*. *Acute dissection superimposed on a pre-existing aneurysm* is considered a life-threatening condition and is therefore another indication for operation.

In the chronic phase, the indications for operative intervention for aortic dissections are similar to those for degenerative thoracic aortic aneurysms. These indications include rapid expansion of the aneurysm and other factors that increase the likelihood of rupture. Elective operation is considered when the affected segment has reached a diameter of 6.0 to 6.5 cm or when an aneurysm has enlarged by >1 cm during a 1-year period. A lower threshold often is used for patients with Marfan syndrome.

Endovascular Treatment. Endovascular therapy (fig. 16) is routinely used in patients with descending aortic dissection complicated by visceral malperfusion. Abdominal malperfusion syndrome often is fatal; prompt identification of visceral ischemia and expedited treatment to restore hepatic, GI, and renal perfusion are imperative for a positive outcome.

In the endovascular technique known as *endovascular fenestration*, a balloon is used to create a tear in the dissection flap, which allows blood to flow in both the true and false lumens. Placement of a stent graft in the true lumen of the aorta can resolve a «dynamic» malperfusion.

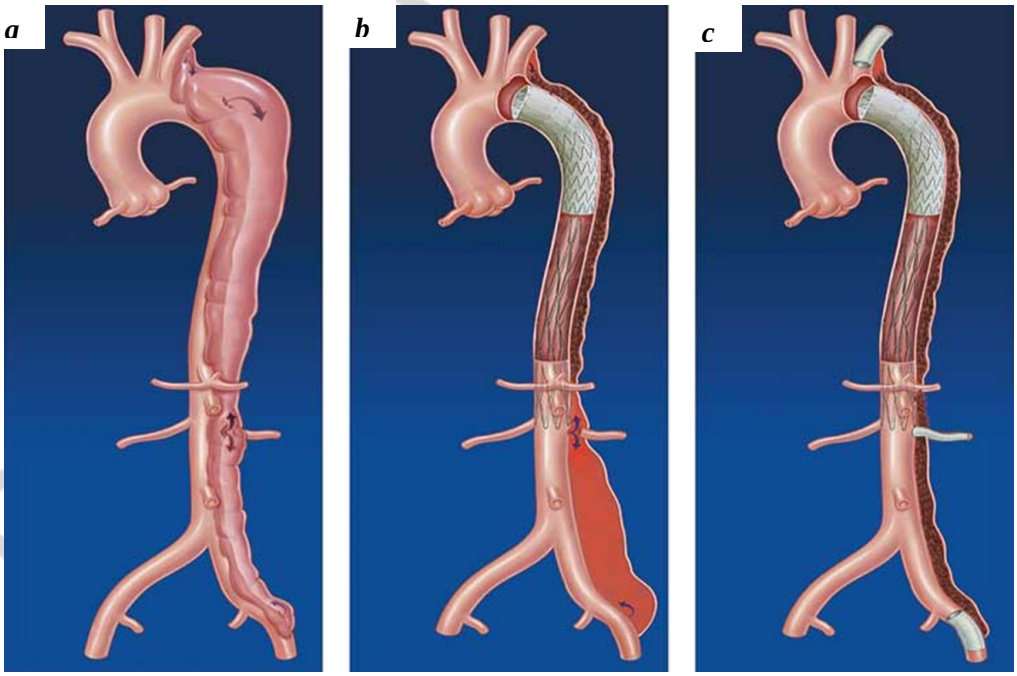


Fig. 16. Steps of endovascular repair of the descending aorta dissection

ABDOMINAL AORTIC ANEURYSM

Abdominal aortic aneurysm (AAA) is a relatively common and often fatal condition that primarily affects older patients. With an aging population, the incidence and prevalence of AAA is certain to rise. Most AAAs are asymptomatic, and physical examination lacks sensitivity for detecting an aneurysm. They can be classified by **localization** (fig. 17) and by **size** (fig. 18). In 95 %, the location of aortic aneurysms is the infrarenal aorta with 18–20 % rate of involving the aortic bifurcation and iliac arteries.

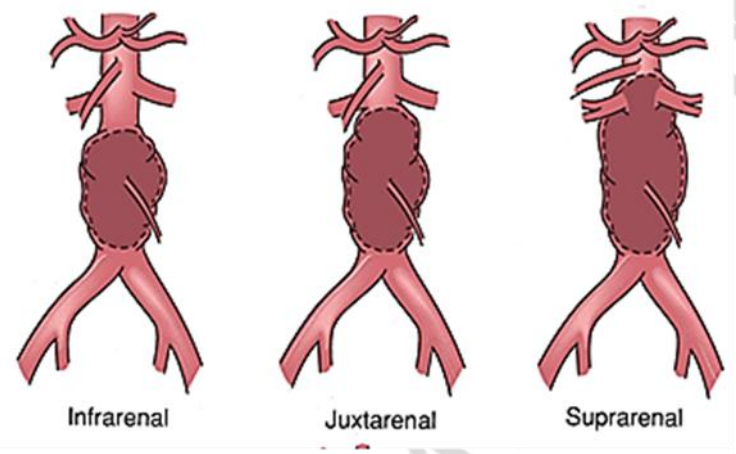


Fig. 17. Classification of AAA according to origin of renal arteries

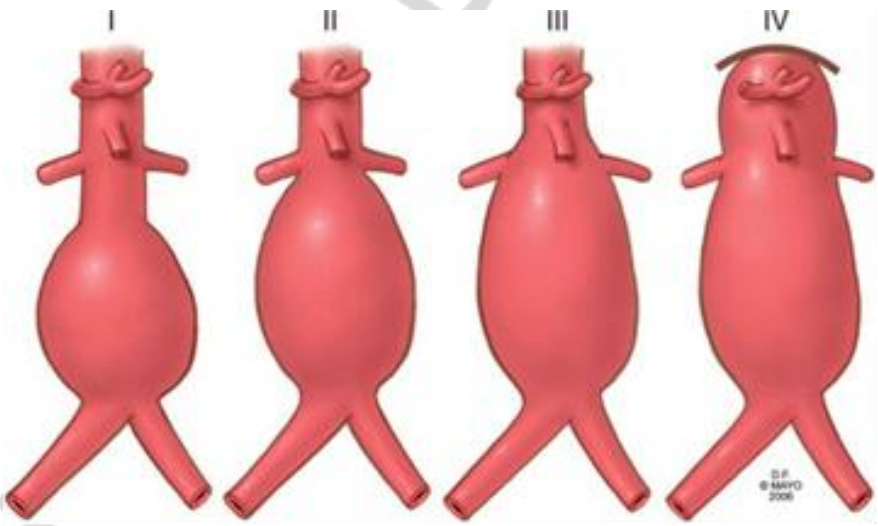


Fig. 18. Four grades of AAA by size

CAUSES AND RISK FACTORS

The pathogenesis of aneurysmal disease of the aorta is complex and multifactorial. A degenerative process in the aortic wall is the most common cause of AAA development. Atherosclerotic disease, age, male sex, smoking history, family history, hypertension, coronary artery disease, and chronic obstructive pulmo-

nary disease are associated with the development of AAA. Diabetes and black race have negative association with AAA. Other less common causes include inflammation, infection, and connective tissue disease.

Inflammatory AAA accounts for 5% to 10% of all AAAs. In contrast to atherosclerotic AAA, the inflammatory variant is characterized pathologically by marked thickening of the aneurysm wall, fibrosis of the adjacent retroperitoneum, and rigid adherence of the adjacent structures to the anterior aneurysm wall.

Male sex and smoking are even stronger risk factors in inflammatory AAA. Smoking cessation is the first step of medical therapy, followed by surgical repair.

Patients with connective tissue disorders such as Marfan's syndrome and Ehlers-Danlos syndrome tend to have more extensive and larger aneurysms at a younger age.

NATURAL HISTORY

The natural history of an AAA is to expand and *rupture* with severe retroperitoneal or abdominal bleeding. Average growth is approximately 3 to 4 mm/year. Rupture risk appears to be directly related to aneurysm size as predicted by Laplace's Law. The annualized risk of rupture is given in tabl. 1. The rupture risk is quite low below 5.5 cm and begins to rise exponentially thereafter. Rapid expansion (or *growth leap*) of > 5 mm within 6 months also increases the risk of rupture. Other complications of AAA include: *thrombosis of an aneurysm, distal embolization,* and *infection*.

Table 1

Annualized risk of rupture of abdominal aortic aneurysm (AAA) based on size

Description	Diameter of aorta, cm	Estimated annual risk of rupture, %	Estimated 5-year risk of rupture, %*
Normal aorta	2–3	0	0 (unless AAA develops)
Small AAA	4–5	1	5–10
Moderate AAA	5–6	2–5	30–40
Large AAA	6–7	3–10	>50
Very large AAA	>7	>10	Approaching 100

*The estimated 5-year risk is more than five times the estimated annual risk because over that 5 years, the AAA, if left untreated, will continue to grow in size

CLINICAL MANIFESTATIONS

Most AAAs are asymptomatic, and they are usually found incidentally during work-up for *chronic back pain* or kidney stones. Physical examination is neither sensitive nor specific except in thin patients. Large aneurysms may be missed in the obese, while normal aortic pulsations may be mistaken for an aneurysm in thin individuals. Rarely, patients present with back pain and/or abdominal pain with a tender pulsatile mass. ***Patients with these symptoms must be treated as if they had a rupture until proven otherwise***.

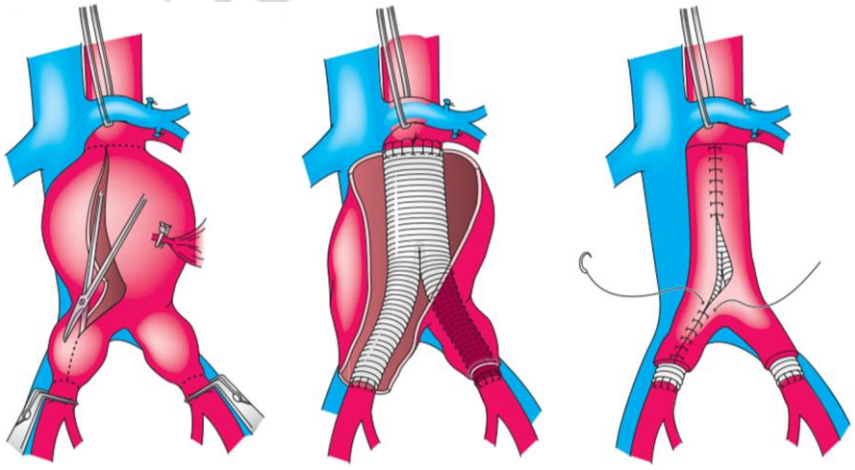
Patients who are hemodynamically unstable with a history of acute back pain and/or syncope, and a known unrepaired AAA or a pulsatile abdominal mass are considered to have an aneurysm rupture and should be immediately taken to the operation room. ***Overall mortality of AAA rupture is 71 to 77 %,*** which includes all out-of-hospital and inhospital deaths. Nearly one half of all patients with ruptured AAA die before reaching the hospital.

TREATMENT

The treatment of abdominal aortic aneurysm represents a balance between the risk of rupture and the operative mortality rate. Infrarenal abdominal aortic aneurysms should be repaired in men when the diameter reaches 5.5 cm and in women when the diameter reaches 5.0 cm provided they are a reasonable operative risk.

Unless symptomatic or ruptured, AAA repair is a prophylactic repair. The rationale for recommending repair is predicated on the assumption that the risk of aneurysm rupture exceeds the combined risk of death from all other causes such as cardiopulmonary disease and cancer. On the other hand, our limitation in predicting timing and cause of death is underscored by the observation that over 25 % of patients who were deemed unfit for surgical repair because of their comorbidities died from rupture of their aneurysms within 5 years.

Open repair. For the repair of AAA the aneurysm sac is opened, and a prosthetic graft is used to reconstruct the aorta. If the aneurysm only involved the abdominal aorta, a linear tube graft can be used to replace the aorta. If the aneurysm extends distally to the iliac arteries, a prosthetic bifurcated graft is used for either an aortobiiliac or aortobifemoral bypass reconstruction (fig. 19). The overlying aneurysm sac and the retroperitoneum are closed to cover the prosthetic bypass graft to minimize potential bowel contact to the graft. Small and large intestines are returned to the abdominal cavity followed by the closure of the abdominal fascia and skin.

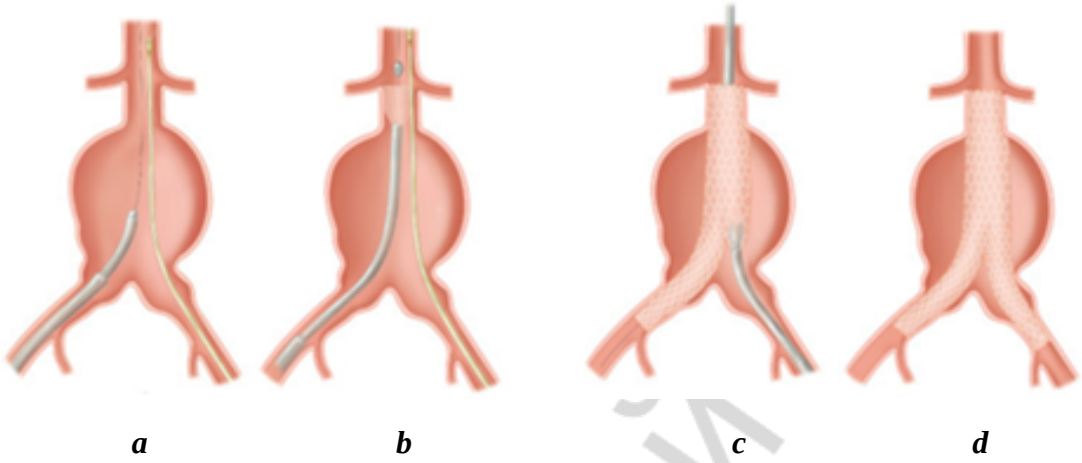


a ***b*** ***c***

Fig. 19. Steps of open repair of AAA

Overall mortality is 2 to 6 % for prophylactic open surgical repair of AAA as compared to 40–50 % for emergent surgery.

Endovascular Repair. The principle of endovascular repair of AAA involves the implantation of an aortic stent graft that is fixed proximally and distally to the nonaneurysmal aortoiliac segment, and thereby endoluminally excludes the aneurysm from the aortic circulation (fig. 20). Unlike open surgical repair, endovascular treatment does not remove or eliminate the aneurysm sac, which therefore is subjected to potential aneurysm expansion or even rupture. The choice of open or endovascular repair is complicated and contingent upon several factors including feasibility, outcome, comorbidities, compliance, cost, and preference.



a ***b*** ***c*** ***d***

Fig. 20. Steps (A-C) of endovascular repair of AAA

COARCTATION OF THE AORTA

Coarctation of the aorta (COA) is defined as a luminal narrowing in the aorta that causes an obstruction to blood flow (fig. 21). This narrowing is most commonly located distal to the left subclavian artery. The embryologic origin of COA is a subject of some controversy. One theory holds that the obstructing shelf, which is largely composed of tissue found within the *ductus arteriosus*, forms as the *ductus arteriosus* involutes. The other theory holds that a diminished aortic isthmus develops secondary to decreased aortic flow in infants with enhanced ductal circulation.

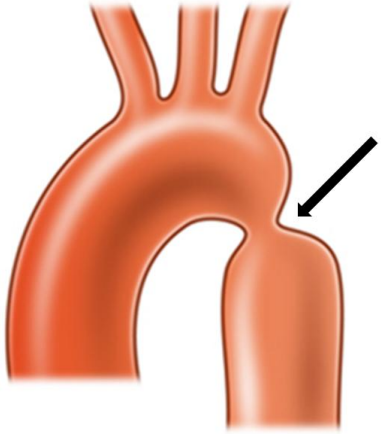


Fig. 21. Aortic coarctation; arrow shows narrowed segment of the aorta

Extensive collateral circulation develops, predominantly involving the intercostals and mammary arteries as a direct result of aortic flow obstruction (fig. 22). This translates into the well-known finding of «rib-notching» on chest radiograph, as well as a prominent pulsation underneath the ribs.

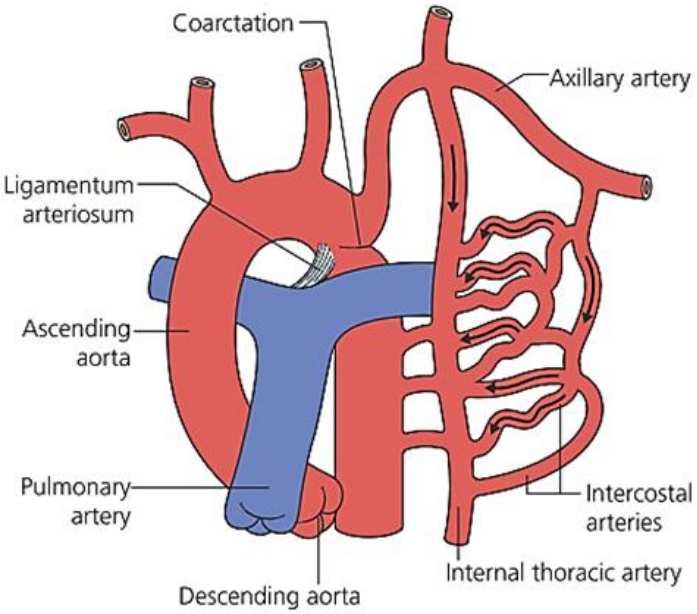


Fig. 22. Paths of collateral circulation

PATHOPHYSIOLOGY

Infants with COA develop symptoms consistent with left ventricular outflow obstruction, including pulmonary overcirculation and, later, biventricular failure. In addition, proximal systemic hypertension develops as a result of mechanical obstruction to ventricular ejection, as well as hypoperfusion-induced activation of the renin–angiotensin–aldosterone system.

DIAGNOSIS

COA is likely to become symptomatic either in the newborn period if other anomalies are present or in the late adolescent period with the onset of left ventricular failure. Physical examination will demonstrate a harsh murmur localized to the left chest and back. Femoral pulses will be dramatically decreased when compared to upper extremity pulses, and differential cyanosis may be apparent until ductal closure.

Echocardiography will reliably demonstrate the narrowed aortic segment, as well as define the pressure gradient across the stenotic segment. Aortography is reserved for those cases in which the echocardiographic findings are equivocal.

THERAPY

The routine management of hemodynamically significant COA in all age groups has traditionally been surgical. The most common surgical techniques in current use are *resection* with end-to-end anastomosis (fig. 23).

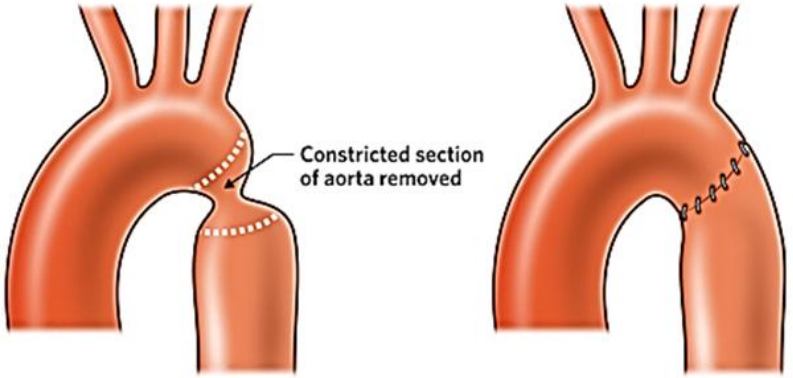


Fig. 23. Surgical resection with end-to-end anastomosis;
a — borders of resection; *b* — the final look

The subclavian flap aortoplasty (fig. 24) is another repair, although it is used less frequently in the modern era because of the risk of late aneurysm formation and possible underdevelopment of the left upper extremity or ischemia. In this method, the left subclavian artery is transected and brought down over the coarcted segment as a vascularized patch. The main benefit of these techniques is that they do not involve the use of prosthetic materials, and evidence suggests that extended end-to-end anastomosis may promote arch growth, especially in infants with the smallest initial aortic arch diameters.

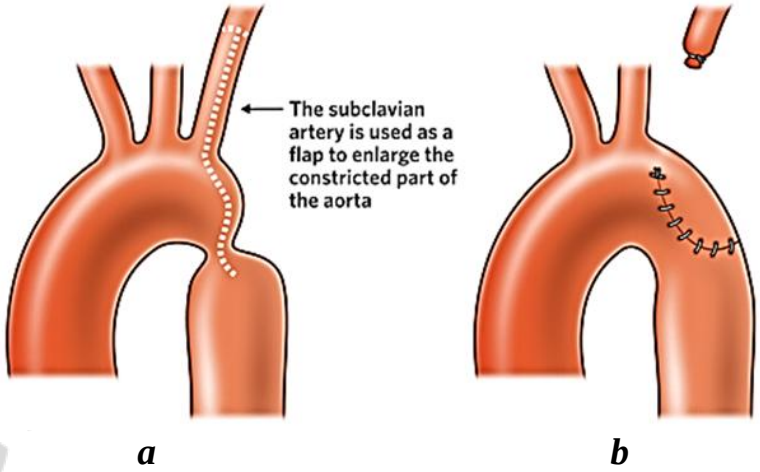


Fig. 24. Subclavian flap aortoplasty;
a — position of the incision; *b* — the final look

End-to-end anastomosis may not be feasible when there is a long segment of coarctation or in the presence of previous surgery, because sufficient mobilization of the aorta above and below the lesion may not be possible. In this instance, pros-

thetic materials, such as a *patch aortoplasty* (fig. 25), in which a prosthetic patch is used to enlarge the coarcted segment, or an *interposition of linear tube graft* must be used. Sometimes, if resection of narrowed segment is contraindicated or very difficult, the bypass surgery (fig. 26) may improve the aortic hemodynamic.

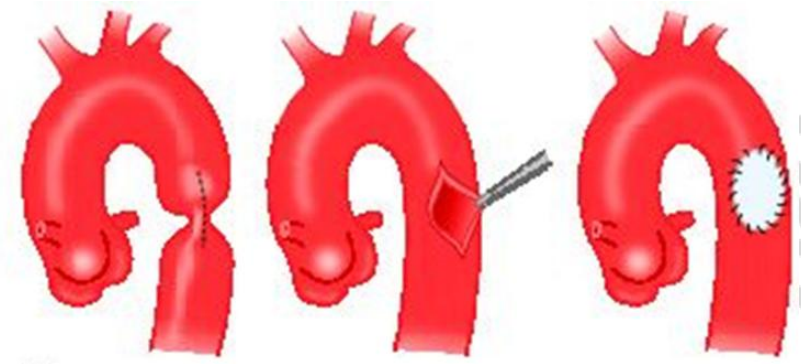


Fig. 25. Aortoplasty with a prosthetic patch

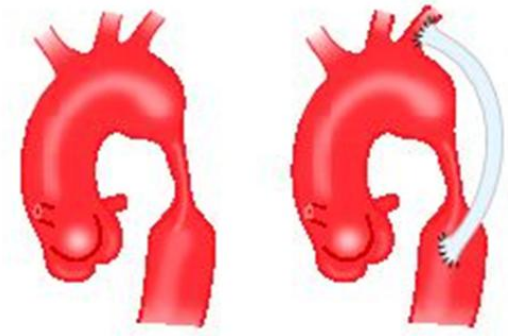


Fig. 26. Surgical bypass

Although operative repair is still the gold standard, treatment of COA by catheter-based intervention has become more widespread. Both balloon dilatation (fig. 27) and primary stent implantation have been used successfully.

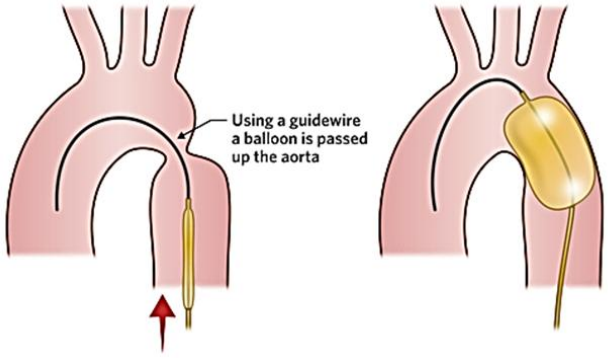


Fig. 27. Balloon dilatation of the narrowed segment

Summary, children younger than age 6 months with native COA should be treated with surgical repair, while those requiring intervention at later ages may be ideal candidates for balloon dilatation or primary stent implantation. Additionally, catheter-based therapy should be used for those cases of restenosis following either surgical or primary endovascular management.

TESTS

1. Parts of thoracic aorta are:

a) Root;
b) Ascending part;
c) Posterior part;
d) Descending part;
e) Intermediate part;
f) Arch;
g) Anterior part.

2. Possible causes of thoracic aortic aneurysms are:

a) Nonspecific medial degeneration;
b) Myocardial infarction;
c) Marfan syndrome;
d) Syphilis;
e) Traumatic pseudoaneurysms;
f) Disease of Fallot;
g) Atrial fibrillation.

3. True aortic aneurysm:

a) The wall has the same structure as aorta;
b) It is the hematoma around ruptured aorta;
c) Also called pseudoaneurysm.

4. Complications of thoracic aortic aneurysm are:

a) Stenosis;
b) Dissection;
c) Rupture;
e) Pulmonary insufficiency;
f) Aortic valve regurgitation;
g) Distal embolization.

5. Signs of aortic dissection are:

a) Acute thrombosis of the aorta;
b) A tear in the intima and inner media of the aorta;
c) Separation of the layers of the aortic media with formation of *false lumen;*
d) Decreasing of aortic diameter;
e) It is associated with aortic aneurysm in 100% of cases.

6. Complications of aortic dissection are:

a) Rupture of the aorta;
b) Myocardial infarction;
c) Acute malperfusion syndrome in coronary, carotid, intercostal, visceral, renal, iliac arteries, etc.;
d) Pulmonary bleeding;
e) Acute aortic valve regurgitation;
f) Formation of abdominal aneurysm.

7. The main cause of abdominal aortic aneurysm is:

a) Trauma;
b) Congenital disorders;
c) Atherosclerosis;
d) Syphilis;

8. Signs on abdominal aortic aneurysm rupture:

a) Hypertension;
b) Acute atrial fibrillation;
c) Acute abdominal or back pain;
d) Hemodynamic instability;
e) Syncope;
f) Absence of pulsation of the abdominal aorta.

Answers: 1 – a, b, d, f; **2** – a, c, d, e; **3** – a; **4** – b, c, e, f; **5** – b, c; **6** – a, c, e; **7** – c; **8** – c, d, e.

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