Chapter

Aortic Dissection

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Abstract

Aortic dissection remains one of the rare but life-threatening causes of chest pain presenting to the emergency department. High index of suspicion is required for prompt diagnosis of the cases presenting to the ED. Symptoms may vary with extent and the progression of the dissection and may further complicate the diagnosis. Thus, patients may present with features of acute MI, CVA, or other end-organ ischemia. Hypertension at presentation may be an important clue for diagnosis of underlying dissection. In low risk patients, D dimer may become a useful screening tool. In patients with high index of suspicion, the choice of investigation will depend on the overall stability of the patient and extent of end-organ ischemia. Stable patients may benefit from CT angiography due to its widespread availability and speed of acquisition. Diagnosis may be challenging for hemodynamically unstable patients in centers where the resources are limited. Transesophageal echocardiography may provide diagnosis in such patients at bedside or in the emergency department. Prompt investigations are required to accurately define the type and extent of damage so that the patient receives life-saving measures in a timely manner.

Keywords: aortic dissection, D dimer, CT angiography, transesophageal echocardiography

1. Classification

Aortic dissection (AD) has been conventionally classified based on anatomical considerations (**Figure 1**).

1.1 Anatomical classification

1.1.1 DeBakey classification

This classification takes into consideration the site of origin of the intimal tear and classifies AD into three types:

- Type I—Originating in the ascending aorta and propagating at least to the aortic arch.
- Type II—Originating and confined to the ascending aorta.
- Type III—Originating in the descending aorta.

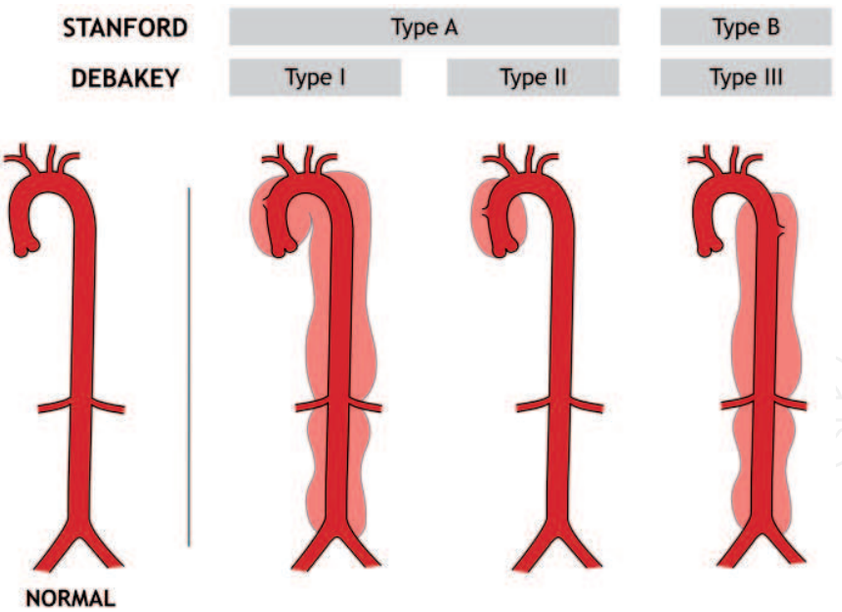

Figure 1.
Anatomical classification of aortic dissection.

1.1.2 Stanford classification

This classification takes into consideration involvement of the ascending aorta irrespective of the origin of the intimal tear:

- Type A—Dissection involves the ascending aorta.
- Type B—Dissection spares the ascending aorta.

Stanford classification is widely used to drive further decision-making and management as any involvement of the ascending aorta (type A) is a surgical emergency, whereas type B can be managed conservatively.

1.2 Etiological classification

Aortic dissection can also be classified based on etiological considerations.

1.2.1 Sporadic

Not associated with any known congenital or genetically mediated syndromes.

1.2.2 Genetically mediated

Associated with known genetic syndromes like Marfan's syndrome, Ehlers-Danlos syndrome, Turner syndrome, and Loeys-Dietz syndrome.

1.2.3 Congenital cardiovascular defects

Bicuspid aortic valve, coarctation of aorta, and annuloaortic ectasia.

1.2.4 Traumatic

Prior cardiac surgery, blunt injury, and cardiac catheterization.

1.3 Newer classification

With advancement of treatment approach in cases previously managed conservatively, especially with endovascular interventions, recent classifications have taken into consideration other factors like, the duration of symptom onset, evolution of complications, and extent of involved segments.

1.3.1 Dissect classification

The DISSECT classification system is a mnemonic-based approach with relevance to the therapeutic considerations, including endovascular management. The six features of aortic dissection include duration of disease, intimal tear location, size of the dissected aorta, segmental extent of aortic involvement, clinical complications of the dissection, and thrombus within the aortic false lumen [1].

1.3.2 Penn ABC classification

The Penn ABC classification further divides type A aortic dissection based on evolution of complications:

- Aa—Absence of branch vessel malperfusion or circulatory collapse
- Ab—Branch vessel malperfusion with ischemia
- Ac—Circulatory collapse with or without cardiac involvement
- Abc—Both branch vessel malperfusion and circulatory collapse (localized and generalized ischemia)

1.3.3 Classification based on duration of symptoms

More recently type B aortic dissection has been classified based on duration of symptom onset [2]:

- Acute—less than 2 weeks
- Subacute—2 weeks to 92 days
- Chronic—>92 days

2. Pathophysiology

The pathophysiology of AD involves the breakdown of the intima and/or the media. The initiating event is an intimal tear. Less commonly rupture of the vasa vasorum may be the initiating event. Initial tear is commonly at the site of greatest hydraulic stress which are the right lateral wall of the ascending aorta in about 50–65% of the cases and the proximal segment of the descending aorta (20–30%) [3]. Subsequently, intramural extension of the bleeding both longitudinally and circumferentially causes the separation of aortic wall layers creating a true lumen and a false lumen. A further intimal tear may create a communication between the false lumen and true lumen. The dissection can extend in antegrade or retrograde

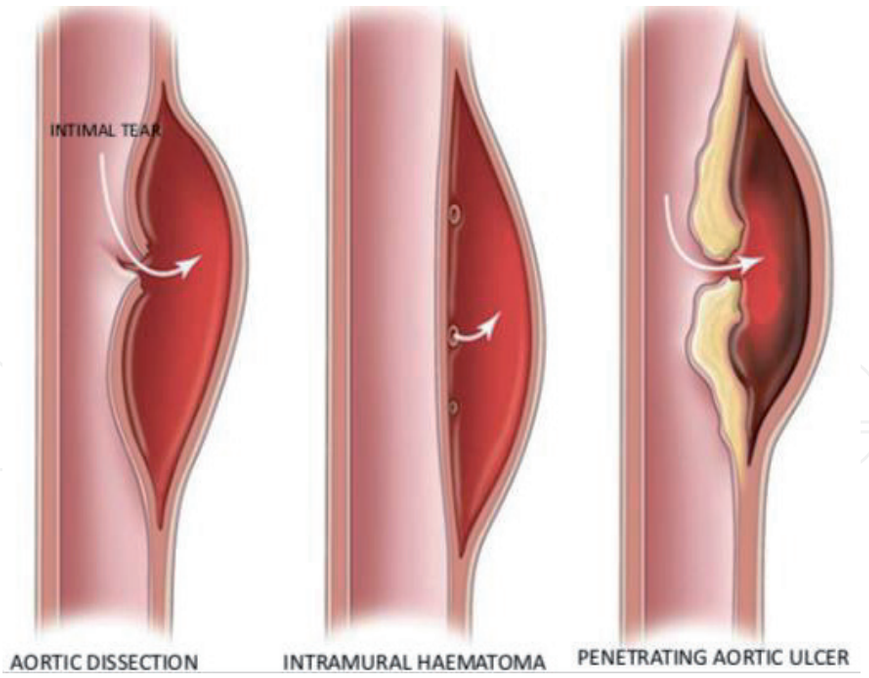

Figure 2.
Acute aortic syndrome.

directions from the site of origin leading to complications including acute aortic insufficiency, cardiac tamponade, and organ ischemia and with disruption of the adventitial layer may lead to aortic rupture.

Intramural hematoma (IMH) is characterized by bleeding confined to the medial layer with no intimal tear visualized by current imaging studies.

Rarely ulceration of an atherosclerotic lesion penetrating to the medial layer may give rise to a penetrating aortic ulcer with similar consequences as of AD.

These three conditions, namely, AD, IMH, and penetrating aortic ulcer, are categorized under the broader category of acute aortic syndrome (AAS) (**Figure 2**) [4].

3. Predisposing conditions

Factors that increase the risk of aortic dissection in a person's life include the following;

3.1 Age and sex

Aortic dissection tends to occur more in men between 60 and 80 years old, whereas women are generally older than men having aortic dissection [5]. However both men and women can develop these conditions at any age, but in females the outcome is worse. On the other hand, familial aortic dissections occur in younger patients as compared to sporadic aortic dissection [6].

3.2 Hypertension

Systemic hypertension is the most important predisposing condition for aortic dissection. It could be either an acute, transient, abrupt rise in blood pressure leading to aortic dissection by various mechanisms like strenuous resistance exercises, weight lifting or illicit use of drugs like cocaine, egotism, and energy drink usage [7, 8]. While chronic or long-term hypertension keeps greater pressure on atherosclerotic arterial walls, leading to intimal tear and aortic aneurysm.

3.3 Genetic disorders

People having specific genetic conditions have a higher incidence of aortic dissection, like Marfan's syndrome, Turners syndrome, Ehlers-Danlos syndrome, annuloaortic ectasia, adult polycystic kidney disease, Noonan syndrome, and osteogenesis imperfecta. Mostly patients with Marfan's syndrome who develop aortic dissection are young, around 40 years old, and have family history of Marfan's syndrome and aortic dissection [9].

3.4 Bicuspid aortic valve

Bicuspid aortic valve usually leads to aortic dissection of ascending aorta, because of severe loss of elastic fibers in the media wall. Patients with bicuspid valves associated with aortic dissection are younger below 40 years age [10].

3.5 Coarctation of aorta

The most common area for congenital coarctation of aorta is the site of ductus arteriosus, where the aorta is focally narrowed. That area is usually underdeveloped, hypoplastic, and small, affecting the layers of the aorta, and increases the risk for aortic dissection.

3.6 Inflammatory or infectious conditions

Inflammatory or infectious diseases that lead to vasculitis (like giant cell arteritis, rheumatoid arthritis, takayasu arteritis, syphilitic aortitis, etc.) affect the vaso vasorum or small arteries that supply blood to the aortic wall [11]. When these small arteries are compromised, they lead to the ischemic injury to the aortic wall and predisposes to aortic dissection. For example, in tertiary syphilis, inflammation begins at adventitia of the aortic arch leading to obliterative endarteritis of the vasa vasorum, luminal narrowing, ischemic injury of medial aortic arch, and finally loss of elastic support and vessel dilatation.

3.7 Blunt chest trauma

The aortic area most commonly involved in blunt chest trauma is the proximal descending aorta, due to its relative mobility over fixed abdominal aorta which is held in place by ligamentum arteriosum. Usually an acute deceleration injury in motor vehicle accident leads to aortic rupture or dissection.

3.8 Aortic instrumentation or previous heart surgery

Cardiac surgery or instrumentation for coronary or valvular heart diseases can be complicated by aortic tear, abnormal dilatation of aorta, and risk for aortic dissection [12].

3.9 Pregnancy and delivery

Both pregnancy and delivery are independent risk factors for aortic dissection [13], but with the presence of other connective tissue diseases like Marfan's syndrome or bicuspid aortic valve, the risk usually multiplies. In pregnancy, aortic dissection occurs most commonly in third trimester due to hyper dynamic metabolic state and hormonal effects on the vasculature.

3.10 Fluoroquinolone usage

Some observational studies relate an increased association of aortic dissection or aneurysm with fluoroquinolone usage [14].

4. Clinical features of aortic dissection

The signs and symptoms of aortic dissection depends upon the extent of dissection and compression of adjacent vascular structures.

4.1 Symptoms

4.1.1 Chest pain

The most common symptom is severe pain of sudden onset, described by patient as sharp stabbing or tearing type. When pain localized to anterior chest wall, neck, or jaw, the point of origin of the aortic dissection is from the ascending aorta, and when it is localized to the interscapular area, abdomen, and back, the descending aorta is usually involved.

Pain that is localized to the abdomen must raise the possibility of involvement of the mesenteric artery.

In few cases the patient may present with pleuritic pain if pericardial hemorrhage occurs.

Dissection may present rarely without pain only and mostly in older patients in cases that involve the ascending aorta. Such patients also have more instances of stroke, heart failure, and syncope.

4.1.2 Syncope

Usually happens in aortic dissection presenting with cardiac tamponade or brachiocephalic vessel involvement and occurs in up to 10% of patients.

4.2 Signs

4.2.1 Hypertension

Seen in 30% of type A and 70% of type B disease.

4.2.2 Hypotension

Seen with ascending aortic dissection and may be due to aortic rupture leading to carotid tamponade (more in females), acute aortic regurgitation, acute MI, hemothorax ,or hemoperitoneum.

4.2.3 Transient pulse deficits

This results from the intimal flap or hematoma blocking or compressing the artery. It is common in dissection involving the aortic arch and thoracic and abdominal aorta.

Patients who presented with pulse deficits had more chances of having hypotension, coma, or neurological deficits. Such patients also had higher rates of complications and mortality.

4.2.4 Cardiac murmurs

Aortic dissection involving the aortic valve results in aortic regurgitation and an early diastolic murmur in the Erb's point (Austin Flint murmur). It occurs in about 50–75% of all ascending aortic dissections.

4.2.5 Focal neurological deficits

Occurs when the aortic dissection involves the proximal branch arteries and compression of adjacent structures. The deficits may be:

- Stroke/altered consciousness
- Horner Syndrome (compression of cervical sympathetic ganglia)
- Hoarseness of voice (compression of the left recurrent laryngeal nerve)
- Acute paraplegic (spinal cord ischemia from intercostal vessel compression)

Also seen are:

- Collapsing pulse
- Wide pulse pressure
- Congestive cardiac failure
- Pleural effusion (commonly left-sided)
- Pulsating neck swelling

5. Work-up/diagnosis

In chronological order as the patient is admitted into ER with a clinical picture suggestive of aortic dissection:

5.1 Electrocardiography

ECG changes may mimic acute cardiac ischemia, which make it further difficult to distinguish aortic dissection from acute myocardial infarction in the presence of chest pain.

If the dissection involves the coronary ostia, the right coronary artery can be affected, which will lead to ST segment elevation in a similar pattern to inferior wall infarction.

In most cases, there will be non-specific ECG changes, or ECG can be normal.

5.2 Blood investigations

CBC: there may be **leukocytosis** due to stress state. **Low hemoglobin** and hematocrit suggests bleeding (dissection is leaking or has ruptured).

Elevated creatinine and BUN may indicate involvement of renal arteries (in such scenario you would expect hematuria, oliguria, or anuria), or it may indicate dehydration due to pre-renal blood loss (dissection is leaking or has ruptured).

Troponin I and T may be elevated if the dissection has involved the coronary arteries and caused myocardial ischemia.

LDH (lactate dehydrogenase) may be elevated due to hemolysis in the false lumen.

D dimer: high negative predictive value. **Aortic dissection is less likely if D dimer is negative** [15].

5.3 Chest radiography

Widening of mediastinum is the classic finding (approximately 60% of cases), but it may not reveal any abnormality. In any case it should not delay the performance of further imaging as CT or MRI [16]. A tortuous aorta (common in hypertensive patients) may be mistaken for widened mediastinum; other differential diagnoses for widened mediastinum include enlarged thyroid, lymphoma, tumors, and adenopathy [17].

Hemothorax is expected to be seen as blood can accumulate in pleural space following dissection rupture.

Ring sign (aortic displacement more than 5 mm past the calcific aortic intima) and abnormal aortic contour can be seen in some patients.

Other radiological abnormalities can be seen including esophageal deviation, tracheal deviation to the right, depression of left mediastinal bronchus, left apical cap, pleural effusion, and loss of paratracheal stripe.

5.4 Echocardiography

Transesophageal echocardiography has higher diagnostic index (sensitivity 99% and specificity 97%) than transthoracic echocardiography (sensitivity 80% and specificity 90%) [18].

TEE is as accurate as CT and MRI, and it can be used at bedside which makes it suitable for hemodynamically unstable patients.

Limitations of transesophageal echocardiography:

- Operator dependent.
- Difficulty in obese patients.
- Not suitable for patients with esophageal stenosis or varicosities.
- Narrow intercostal space, pulmonary emphysema, and mechanical ventilation decrease its accuracy.
- Upper ascending aorta and arch may not be evaluated well.
- False-positive results may occur due to reverberations in the ascending aorta [19].

5.5 Computed tomography

CT with contrast is used more frequently in emergency department settings, only on hemodynamically stable patients who do not have adverse reaction to the intravenous contrast agents.

A 2014 guideline form American College of Radiology recommends CT angiography as the definite modality if there is high clinical suspicion for aortic dissection [16].

CTA provides detailed anatomic definition of the dissection and information about plaque formation.

Spiral (helical) CT is associated with higher rate of detection and better resolution than incremental CT scanning. Imaging information, including type of the lesion, location of pathologic lesion, extent of the disease, and evaluation of the true and false lumen can be assessed quickly and help the surgeon plan the operation [16, 20].

Limitations of CT:

- Cannot provide information about aortic regurgitation.
- CTA not suitable for patient with renal impairment or allergy to contrast material.
- Hemodynamic unstable patients cannot be shifted to radiology department.

5.6 Smooth-muscle myosin heavy-chain assay

Performed in the first 24 hours. **Levels are higher in the first 3 hours**; a 2.5-fold increase has a sensitivity of 91% and specificity of 98% for aortic dissection.

5.7 Measurement of the degradation products of plasma fibrin and fibrinogen

Plasma fibrin degradation product level (FDP) of **12.6 μg/mL or higher** is suggestive of the possibility of aortic dissection with false lumen in symptomatic patient.

Plasma fibrin degradation product level (FDP) of 5.6 μg/mL or higher is suggestive of the possibility of dissection with complete thrombosis of the false lumen [21].

5.8 Magnetic resonance imaging (MRI)

MRI has 98% sensitivity and specificity in detection of thoracic aortic dissection.

MRI shows the site on intimal tear, type and extent of dissection, presence of aortic insufficiency, as well as surrounding mediastinal structures. It has the advantage of not using contrast material; thus, it is preferred in patients with renal impairment or allergic to iodine [22].

Limitations of MRI:

- Not suitable for hemodynamically unstable patients.
- Requires much more time than CT.
- Not suitable for patients with pacemaker and other metallic implants [23].

5.9 Aortography

5.9.1 The gold-standard diagnostic modality for aortic dissection

Benefits include accurate visualization of the true and false lumen, intimal flap, aortic regurgitation, and coronary arteries.

Limits of aortography:

- Invasive procedure.

- Not suitable for patient with renal impairment or allergic to contrast.
- Not suitable for hemodynamically unstable patients.
- The false lumen and intimal flap may not be visualized if the false channel is thrombosed.
- Simultaneous opacification of the true and false lumen may mask the dissection.

5.9.2 Diagnosis

Diagnosis is usually done through imaging. The choice of the imaging technique depends on the patient condition (whether he is hemodynamically stable or not).

Chest radiography is the initial basic imaging technique, but it may reveal no abnormality.

Further imaging options like computed tomography (CT) and CT angiography with three-dimensional reconstruction are of higher diagnostic value.

Magnetic resonance imaging (MRI) is as accurate as CT and may benefit patients who have adverse reaction to intravenous drug agents.

In hemodynamically unstable patient, echocardiography is ideal.

Aortography is the gold-standard diagnostic modality.

6. Differential diagnosis

6.1 Myocardial infarction

Typically presents with severe substernal or left-sided chest discomfort radiating to shoulders or left arm and shortness of breath and can be differentiated from AD by the typical ECG changes and the rise in the cardiac markers.

6.2 Myocarditis

Viral myocarditis is often preceded by flu-like symptoms, fever, joint pain, or features of upper respiratory tract infection. These patients usually present with heart failure, and ECHO is done to exclude it from other causes of heart failure.

6.3 Pericarditis or cardiac tamponade

Presents with sharp chest pain and may have a pericardial friction rub. Patients with tamponade present with cardiogenic shock and have low-voltage ECG with electrical alternans and enlarged cardiac shadow on the Chest X-ray.

6.4 Pulmonary embolism

Classically presents with sudden onset of chest pain, shortness of breath, and hypoxia. In patients suspected to have pulmonary embolism, CT pulmonary angiogram is the definitive investigation to establish the diagnosis.

6.5 Tension pneumothorax

Patients present with sudden onset of sharp chest pain and desaturation with absent breath sounds. Diagnosis can be established by Chest X-ray.

6.6 Esophageal rupture

Often preceded by history of forceful vomiting, upper gastrointestinal endoscopy or instrumentation. Chest X-ray shows pneumomediastinum, pneumothorax, or pleural effusion.

7. Management of aortic dissection

7.1 Acute management of aortic dissection involves immediate resuscitation

- Intensive blood pressure monitoring preferably with arterial line to maintain SBP between 100 and 120 mmHg and heart rate < 60/min, to prevent the dissection from expanding. This lowering of blood pressure can be attained with:

    - First line—Beta-blockade using labetalol (20 mg iv initially, followed by either 20–80 mg iv boluses every 10 min to a maximal dose of 300 mg or an infusion of 0.5–2 mg/min IV), esmolol (250–500 mcg/kg IV loading dose; then infuse at 25–50 mcg/kg/min, and titrate to maximum dose of 300 mcg/kg/min).

    - Second line—In patients with asthma, allergy, or any contraindication to beta-blockade, calcium channel blockers diltiazem and verapamil can be used.

    - Third line—Vasodilator therapy. If blood pressure remains above 120mmHg and heart rate < 60/min, nitroprusside infusion (0.25–0.5 mcg/kg/min titrated to a maximum of 10 mcg/kg/min) can be initiated. This vasodilator therapy should not be used without first lowering heart rate with beta-/calcium channel blocker.

    - Pain control using iv opioids (tramadol, morphine, fentanyl)

    - Specific management depends on site of dissection

7.1.1 Acute type A dissection

Acute type A dissection is a surgical emergency with a mortality of 1–2% per hour, as these patients are at high risk of complications such as aortic regurgitation, tamponade, myocardial infarction due to compression of the coronary ostium, stroke, and aortic rupture [24].

This excludes patients with significant comorbidities including prior debilitating stroke, ischemic heart disease, renal failure, malignancy, advancing age, and hemorrhagic stroke, which are associated with a bad prognosis.

These patients should also be assessed for any underlying coronary artery disease or any aortic valve disease by intraoperative transesophageal ECHO to identify any wall motion abnormality or aortic valvular defect.

In the International Registry of Aortic Dissection (IRAD) review of 547 patients in which 80 percent of type A patients were treated surgically and the remaining 20 percent were treatment medically, inpatient mortality rates were 27 and 56 percent for surgical and medically treated patients. Medically treated patients were those with advanced comorbidities and aged individuals with poor prognosis [25].

Poor prognostic factors predicting increased mortality in type A patients according to the IRAD study included advanced age, tamponade at presentation, prior

myocardial infarction, prior stroke, ischemia involving kidney or other viscera, advance renal or lung disease, and previous aortic valve replacement [25–31].

Open surgical repair for type A patients involves resection of the dissecting aneurysm and removal of intimal tear, closure of false lumen and repair of aorta using synthetic graft, and aortic valve repair/replacement. Repair of the aortic arch may also be needed depending on the extent of the pathology.

Patients with genetic disease like Marfan causing Aortic regurgitation, bicuspid aortic valve or aortitis need aortic valve replacement [32].

An alternative to open surgical repair in type A patients with ischemic complications like renal, mesenteric, and peripheral ischemia is endovascular stent grafting.

A novel approach involves the hybrid repair of type A dissection with "frozen elephant trunk repair technique," which involves open surgical repair of the ascending aorta in the form of a traditional elephant trunk and endoscopic stent grafting to repair the descending aorta.

Studies have compared the total arch replacement using the frozen elephant trunk repair technique (FET) with the hemi-arch replacement (AHR) for the type A ascending aortic dissection in which the survival for the patients after 5 years was 95.3% for the FET group and 69.0% for the AHR group, indicating that FET techniques prevent further operations for the complications because of the false lumen [25, 33–35].

7.1.2 Type B aortic dissection

Medical management is preferred for uncomplicated cases of type B aortic dissections unless the dissection or aneurysm expands, ischemic complications or aortic rupture occurs, or the patient has persistent uncontrolled hypertension or chest pain, when surgical treatment or endovascular grafting is to be considered.

Conservative treatment for type B patients involves optimal BP control and long-term surveillance with imaging.

In the IRAD study of 384 patients with type B aortic dissection [36], 73 percent of patients were treated medically with mortality rate of 13% within the first week of admission. Factors associated with increased mortality were shock on presentation, widened mediastinum, excessively dilated aorta (≥6 cm), periaortic hematoma, patients with coma or altered consciousness, mesenteric or limb ischemia, acute renal failure, and patients who were treated surgically.

Endovascular stent grafting is done with the stent covering the dissection leading to thrombosis causing closure of false lumen.

Open surgical repair is rarely done in type B patients. It may be needed in those patients with genetic condition like Marfan's syndrome in whom endovascular repair is difficult.

Several trials have compared medical management with endovascular stent grafting in uncomplicated patients with type B aortic dissection demonstrating that at 2 years there is no difference in survival in either of the endovascular versus medical groups (89% versus 96%) [37]; however at 5 years the occurrence of the aortic complications is reduced in the endovascular group improving late outcome [38].

7.2 Long-term management

Optimal blood pressure control is needed to prevent recurrence or aneurysm formation. This is best achieved by oral combination antihypertensive therapy often including oral beta-blockers. Target blood pressure of less than 120/80 mmHg is preferred.

Screening of first-degree relatives should be performed with transthoracic ECHO (TTE) to look for aortic aneurysm.

At discharge transesophageal ECHO (TEE), magnetic resonance imaging (MRA), or CT angiography should be performed to detect for any leak, and serial images should be done at 3, 6, and 12 months to look for any recurrence, expansion, aneurysmal formation, or leak. MRA is preferred to TEE as is it noninvasive. In patients with renal impairment, MRI can be done without gadolinium.

8. Prognosis

8.1 Type a aortic dissection patients

Untreated patients with type A dissections have a mortality rate of 1–2% per hour because of its association with high risk of complications of aortic rupture, tamponade, aortic regurgitation, ischemic complications of myocardial infarction, and stroke [24].

According to the IRAD follow-up study of 303 patients with type A dissection who were discharged from the hospital, surgically treated patients were 273 (90.1%), and medically treated patients were 30 (9.9%). Surgically treated patients had a 1-year survival of 96.1 ± 2.4% and 3-year survival of 90.5 + 3.9%, whereas the medically treated type A dissection patients had a 1-year survival of 88.6 + 12.2% and 3-year survival of 68.7 + 19.8% [39].

8.2 Type B aortic dissection

Uncomplicated type B patients have an overall survival rate of 90% with immediate medical management with effective control of blood pressure [36].

However in type B patients with complications of aortic rupture, expansion of dissection or ischemic complications of organ hypoperfusion the mortality rates were high. According to the IRAD study, the overall inhospital mortality rate was 13% during the first week, for all type B aortic dissection patients. Mortality rate in type B dissection patients with complications, undergoing surgical management, was 32.1%, whereas in those treated with medical management alone, the mortality rate was 9.6% and was 6.5% for those treated with endovascular approach [36].

The 3-year survival rate for type B patients who were discharged from the hospital according to the IRAD registry was 77.6 ± 6.6% for medically treated patients, 82.8 ± 18.9% for surgically treated patients, and 76.2 ± 25.2% for those treated with endovascular approach [40].

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