Chapter 3.4.1 Pathophysiology

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Part of "Chapter 3.4 - Aortic dissection"

Key messages

- Dissection is most common in hypertensive males in their sixth and seventh decades.
- Nearly two-thirds of dissections arise in the ascending aorta.
- Without surgery, 40 per cent of patients with ascending aortic dissections will die within 24 h and only 30 per cent will survive for 1 week.

Introduction

Dissection of the aorta occurs when blood enters the aortic media and dissects the inner from the outer layers. There is usually a discrete proximal intimal tear, and there may be one or more re-entry tears distally. Dissections are regarded as acute during the first 14 days.

Anatomical considerations

The aortic media is composed of a series of concentric elastic lamellar plates, separated by smooth muscle and connective tissue. During development, in response to radial (compressive) and circumferential (tensile) stresses which are maximal at the intima and weakest at the adventitia, the inner lamellas thicken and smooth muscle cells develop more cell–cell contacts. Because of this structural modification, most of the stress-resistant properties are found in the inner two-thirds of the aortic media. There is a clear change at the junction between the middle and outer thirds of the media; this represents an area of weakness where tearing might be expected with exposure to high levels of stress and coincides with the observed plane of most aortic dissections.

The intimal tear is found most frequently approximately 4 cm above the aortic valve in the ascending aorta or just distal to the subclavian artery. The dissection usually propagates distally, but proximal extension from the ascending aorta may involve the aortic valve annulus, and tears distal to the subclavian artery may propagate in both directions to involve the aortic arch and ascending aorta as well as the descending aorta. The false channel usually extends 50 to 67 per cent around the aortic circumference, rarely involving the whole aortic wall. The outer wall of the false channel is thin and may rupture, particularly into the pericardium where the ascending aorta is unsupported, the pleural space (usually the left), or the abdomen. In the absence of frank rupture, blood may extravasate to form mediastinal or pericardial hematomas. In patients who survive the acute event, the false channel tends to become aneurysmal and may rupture subsequently.

Occasionally, the channel may become thrombosed spontaneously. In a minority of patients there is a discrete re-entry tear so that the false channel produces a parallel aortic circulation, albeit not in communication with the aortic branch arteries.

Although an intimal tear is a universal finding in autopsy series, it may be difficult to identify clinically. The events leading to the intimal tear are poorly understood. It is uncertain whether the intimal tear is the primary event, or whether dissection is initiated by bleeding into the media from the vasa vasorum and the intimal tear follows. The major predisposing factors to dissection appear to be the level of shear stress to which the aorta is subjected and, in a proportion of cases, medial degeneration and structural abnormalities of the aortic wall.

With advancing age, elastin fragmentation, fibrosis, and medial necrosis lead to degeneration of the aortic lamellas. These changes are most pronounced in the ascending aorta and become less prominent as the aorta descends into the abdomen. These changes occur as part of the normal aging process but are more prominent in hypertensive patients. Pre-existing dilation of the aorta (as occurs in Marfan's syndrome) may also be relevant. Laplace's law relates wall stress to pressure and diameter: as the aorta dilates, so wall stress increases. Medial degeneration may result from structural abnormalities (e.g. Marfan's and Ehlers–Danlos syndromes) in which dissection may occur without pre-existing hypertension or, possibly, from ischemic necrosis due to occlusion of the vasa vasorum.

A large proportion of dissections take place in aortas without evidence of excessive degeneration. This is true particularly in patients where dissection follows instrumentation of the aorta. It appears that, whereas abnormalities of the aortic wall may make dissection more likely, once the intima and underlying media are damaged, dissections propagate rapidly due to entry of the blood into the media under pressure.

Predisposing causes

The majority of patients with dissection are hypertensive males between the ages of 40 and 70, usually in their sixth or seventh decades. It is more common in the black population than in Caucasians and is relatively rare in orientals. It is more common in Marfan's syndrome and occurs with increased frequency in Ehlers–Danlos, Turner's, and Noonan's syndromes and in patients with bicuspid aortic valves. Such patients tend to develop dissection at a younger age and frequently have no history of arterial hypertension. Rarer associations include pregnancy (usually in the third trimester),

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vasculitis, and aortic coarctation. Dissection may occasionally follow trauma, aortic instrumentation, cocaine abuse, or aortotomy. Syphilis and atheroma lead to saccular aneurysms rather than dissection.

Site and classification of dissection

Sixty to sixty-five per cent of dissections originate in the ascending aorta, 5 to 10 per cent in the aortic arch, and 30 to 35 per cent in the first part of the descending aorta. Subsequent dissection usually occurs distally but may also extend proximally. The original De Bakey classification divides dissection into three types based on the extent of aortic

involvement (not the site of the intimal tear, which may be difficult to identify) (Fig. 1).

Fig. 1 Classification of aortic dissection.

I.   This involves both the ascending and descending aorta and usually extends into the abdomen. Ten to fifteen per cent of such patients have an intimal tear distal to the left subclavian artery; in the remainder the origin is in the proximal aorta.

II. This is the least common type and involves the ascending aorta only, stopping just proximal to the innominate artery. This type of dissection is seen in Marfan's syndrome and may occasionally be found incidentally in operations for ascending aortic aneurysms.

III. This originates distal to the left subclavian artery and extends downwards. Type IIIa dissections are confined to the descending thoracic aorta only, whereas the more common type IIIb dissection extends into the abdominal aorta and may involve the iliac arteries. The majority of these aneurysms have an intimal tear just distal to the origin of the left subclavian artery. Type III dissections tend to occur in older patients.

The alternative Shumway (Stamford) classification divides dissection into two types only. This classification is less descriptive but is well suited to clinical decision-making (Fig. 1).

Type A. Involvement of the ascending aorta (De Bakey types I and II).

Type B. No involvement of the ascending aorta (De Bakey type III).

In autopsy series, type A dissections predominate in a ratio of approximately 2 to 1. In most clinical series, types A and B appear almost equally common, probably because type A dissections are more rapidly fatal and may pass unrecognized.

Consequences of dissection

The acute sequelae of dissection include aortic rupture (usually into the pericardium or pleura), disruption of the aortic valve apparatus leading to acute aortic regurgitation and left ventricular failure, and extension of the dissection into the branch walls of arteries originating from the aorta, leading to distal ischemia or infarction. This is more common in large arteries such as the innominate, subclavian, carotid, or renal arteries, but any artery may be affected. Involvement of intercostal arteries in descending aortic dissections may prejudice blood flow to the anterior spinal artery. Distal extension of type IIIb dissection involves the left iliac artery more commonly than the right. Involvement of branch arteries may lead to ischemia or infarction of the territory supplied.

Natural history following dissection

Without surgery, approximately 40 per cent of ascending aortic dissections are fatal within 24 h and only 30 per cent survive 1 week; 86 per cent of these deaths are due to aortic rupture. Ten per cent of patients will survive for 3 months. Survival is higher in dissections limited to the descending aorta: 10 to 20 per cent will die within the first 24 to 48 h, 75 per cent will survive for 1 month, and over 60 per

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cent will survive for 1 year. Descending aortic dissection tends to occur in an older age group, and much of the mortality is not directly related to the dissection.

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