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The Diagnosis and Management of Aortic Dissection

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Abstract

Aortic dissection represents the most common aortic emergency, affecting 3 to 4 per 100 000 people per year and is still associated with a high mortality. Twenty percent of the patients with aortic dissection die before reaching hospital and 30% die during hospital admission. Aortic dissections may be classified in 3 ways: according to their anatomical extent (the Stanford or DeBakey systems), according to the time from onset (acute or chronic), and according to the underlying pathology (the European Society of Cardiologists' system). Advances in endovascular technology have provided new treatment options. Hybrid endovascular and conventional open surgical repair represent the mainstay of treatment for acute type A dissection. Medical management remains the gold standard for acute and uncomplicated chronic type B dissection, though endovascular surgery offers exciting potential in the management of complicated type B dissection through sealing of the intimal entry tear.

Keywords

aortic dissection, acute aortic syndrome, endovascular

Introduction

The clinical features of aortic dissection were first described in 1761, by Morgagni, who ordered his patient to ''think seriously and piously of his departure from this mortal life, which was very near at hand and inevitable.''[1] Aortic dissection represents the most common aortic emergency, affecting 3 to 4 per 100 000 people per year,[2-4] and is still associated with a high mortality as acknowledged by Morgagni over 200 years ago. Contemporary Swedish registry data show that 20% of patients with aortic dissection died before reaching hospital and 30% died during hospital admission.[4]Circadian and seasonal variation in prevalence has been reported, with peaks of frequency in the morning and in winter.[5,6]

Definition, Pathogenesis, and the Acute Aortic Syndrome

Aortic dissection occurs following separation of the layers constituting the wall of the aorta. The intima and inner media are separated from the outer media and adventitia of the aorta to create a false lumen. Classical ''communicating'' aortic dissection is initiated by an entry tear in the intima, sending pulsatile blood flow into this false passage, which is separated from the true aortic lumen by an intimal flap. Propagation of the false lumen can occur proximally as well as distally and additional tears in the dissecting membrane are termed ''reentry'' sites. ''Noncommunicating'' aortic dissection is a rarer variant characterized by the absence of flow in the false lumen, thought to arise from spontaneous rupture of the aortic vasa vasorum, leading to intramural hematoma.[7] Although classically regarded as a more stable variant of aortic dissection and a precursor of communicating aortic dissection, recent evidence suggests that intramural hematoma has similarly adverse outcomes to classical communicating aortic dissection.[7] High-resolution imaging has identified a range of intramural and intimal pathology thought to be related to aortic dissection, including penetrating atherosclerotic aortic ulcers.[8] The term ''acute aortic syndrome'' has been coined to represent classical aortic dissection and its noncommunicating variants of intramural hematoma and penetrating aortic ulcer; the clinical presentation of these distinct pathological entities may be indistinguishable.

A wide range of risk factors may predispose to aortic dissection through acquired and congenital weakness of the aortic media or intimal disease. Hypertension, increasing age, and atherosclerosis are common comorbidities from the Internation Registry of Acute Aortic Dissection (IRAD) studies.[9-11] Important risk factors include Marfan syndrome with deficiency of

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fibrillin-1,[12] Ehler-Danlos type IV with defective synthesis of type III procollagen,[13] and other connective tissue disorders inducing cystic medial necrosis.[14] Other risk factors include a congenitally bicuspid aortic valve,[15] familial aneurysmal syndromes,[16] aortic vasculitic diseases, cocaine abuse,[11] and pregnancy.[10]

Classification

Aortic dissections may be classified in 3 ways: according to their anatomical extent (the Stanford or DeBakey systems), according to the time from onset (acute or chronic), and according to the underlying pathology (the European Society of Cardiologists' system). The most common classifications in clinical practice are the Stanford and DeBakey systems. The Stanford classification distinguishes between type A dissections involving the ascending aorta and type B dissections, which only involve the descending aorta with the entry tear usually found distal to the ostium of the left subclavian artery.[17] The DeBakey classification has 3 groups; type I involves both ascending and descending aorta, type II involves ascending aorta only, and type III is restricted to the descending aorta.[18] The European Society of Cardiologists classified aortic dissection according to pathogenesis into 5 groups, with implications for endovascular treatment.[19] Type 1 describes classical aortic dissection with an intimal flap separating the true and false lumen. Type 2 involves medial disruption by intramural hematoma. Type 3 is a ''discrete or subtle'' dissection without hematoma, with an eccentric bulge at the entry site. Type 4 is a penetrating aortic ulcer and Type 5 is traumatic or iatrogenic dissection. Types 3, 4, and 5 feature discrete entry points and are theoretically suited to endovascular treatment, which may be prohibited in type 1 by the presence of multiple entry tears and in type 2 by the lack of a clearly defined entry point. Dissections are considered acute for the first 14 days following the intimal entry tear and chronic after 14 days, with important implications for management and survival.

Clinical Presentation and Investigation

Aortic dissection typically occurs following the abrupt onset of sharp chest pain in a male patient in his 60s, worst at the moment of onset[20-22] although up to 10% of patients with aortic dissection do not complain of pain.[22] The pain may radiate to the neck in ascending aortic dissection or to the interscapular area in descending thoracic dissection. Aortic regurgitation and pulse deficit are seen in 31.6% and 15.1% of patients, respectively.[22] Hypotension is seen in 25% of patients with type A dissection, whereas hypertension is typical in type B dissection.[22] The sensitivity and specificity of chest radiographs, electrocardiogram (ECG), or blood biomarkers such as smooth-muscle myosin heavy-chain protein are insufficient to establish or exclude the diagnosis.[23] Spiral computed tomography angiography (CTA), transesophageal echocardiography (TOE), and magnetic resonance imaging angiography (MRA) are the main diagnostic imaging modalities of use, with CTA representing the initial investigation in 61% of cases reported by IRAD.[19] All 3 modalities have sensitivity and specificity in excess of 95%.[24] Spiral computed tomography angiography is more readily available than MRA and allows imaging of the entire aorta, including the relationships of true and false lumens to aortic branch vessels, which facilitates planning of open or endovascular repair. Transesophageal echocardiography allows identification of type A dissections, detection of aortic regurgitation or pericardial effusion, and useful intraoperative assessment of whether the operator is within the true lumen. Its disadvantages include the inability to assess the abdominal aorta. In the acute setting, CTA is the recommended first-line investigation, with MRA reserved for the assessment of stable patients with chronic dissection.[19] Electrocardiogram-gated CT is of particular interest for the differential diagnosis of acute chest pain; it accurately differentiates aortic dissection from coronary atherosclerosis and pulmonary emboli.[25]

Treatment

There is little evidence from randomized controlled trials to guide the optimal management of aortic dissection. The evidence base comprises registry data and large series; best practice is therefore guided by expert consensus.[19,26]

Acute Type A Dissection

The natural history of acute type A dissections mandates surgical intervention; if left untreated, up to 91% of patients will be dead within 1 week.[22] Surgical intervention aims to replace the ascending aorta and arch affected by the intimal dissection with prosthetic graft. It carries an in-hospital mortality of 24%.[27] A broad variety of specific operative techniques may be required to combat the myriad morphological possibilities for type A dissection. Proximal extension to the aortic valve or ostia of the coronary arteries may require replacement of the aortic valve or coronary artery bypass; these techniques were reported in 24% and 15% of IRAD type A dissections, respectively.[27] Distal extension of the dissection beyond the replaced ascending aorta may require a hybrid endovascular procedure to seal the distal intimal tear; the optimal timing of this approach remains controversial and some experts prefer a staged approach.[28-30]

Acute Type B Dissection

In contrast to acute type A dissection, the natural history of uncomplicated type B dissection demonstrates that 40% of patients survive to 1 year without treatment[31]; medical treatment confers an in-hospital mortality of 1% to 11%.[14] The presence of complications such as aortic branch involvement leading to visceral or limb ischemia, rupture, or more diffuse end points such as refractory pain or uncontrollable hypertension requires surgical or endovascular intervention.[19-26] Despite early reports in favor of endovascular stenting for acute type B dissection,[32] a recent expert consensus document has

concluded that in the absence of these complications the medical management of uncomplicated type B dissection provides a satisfactory survival benchmark that is not surpassed by endovascular intervention.[26] Medical treatment comprises observation in an intensive care setting, with tight control of systolic blood pressure between 100 and 120 mm Hg to minimize hemodynamic shear stress and discourage rupture.[23] β-Blockade with propranolol, metoprolol, esmolol, or labetalol is preferred, with sodium nitroprusside used as a second-line agent; patients with chronic obstructive pulmonary disease are given diltiazem or verapamil.[19] Conversely, patients with complications in acute type B dissection have a poor prognosis of 50% mortality and require surgical intervention. Despite open or endovascular treatment, a third of these patients still die.[33]

A meta-analysis of retrospective case series in a total of 609 patients showed a 30-day mortality of 9.8% ± 2.2%, following endovascular repair of acute type B dissection,[34] although significant selection bias should be assumed. Long-term outcomes in endovascular repair of complicated acute type B dissection are poor. False lumen thrombosis may only be achieved in 44% of cases, while 20% rupture within 5 years due to ongoing aortic expansion.[35] Even following false lumen thrombosis of the stented aorta, 16% of patients develop distal perfusion of the false lumen in unstented aorta and many require surgical re-intervention.[35-37] Noncommunicating intramural hematoma or penetrating ulcers should be treated depending on their location in an analogous manner to type A classical dissection which is treated operatively and type B classical dissection treated conservatively.[8]

Chronic Dissection

The optimal management of chronic type B dissection remains unclear. The INSTEAD trial compared optimal medical management with thoracic endovascular aneurysm repair (TEVAR) in type B dissection. There was no difference in all-cause mortality at 2 years (95.6% ± 2.5% with optimal medical therapy; 88.9% ± 3.7% with TEVAR; $P = .15$) and no difference in aortic-related mortality at 2 years ($P = .44$), despite evidence of improved restoration of true lumen with false lumen thrombosis following TEVAR (91.3% with TEVAR vs 19.4% with medical therapy; $P < .001$). Traditional open surgical management of type B dissection has many periprocedural disadvantages compared to TEVAR including the need for posterolateral thoracotomy, single-lung ventilation, cardiopulmonary bypass, hypothermia, heparinization, cerebrospinal fluid drainage, and circulatory arrest to prevent stroke and paraplegia. One year survival after open repair has been reported at 79% with 35% 10-year survival.[38] However, significant reporting bias exists in the available literature. In the absence of compelling level 1 evidence, medical management remains the gold standard for chronic dissection and uncomplicated acute type B dissection.

Follow-Up

Evidence from patients with Marfan syndrome has led to long-term β-blockade for patients with aortic dissection to maintain blood pressure below 135/80 mm Hg.[39] False lumen flow is predictive of aortic expansion and subsequent rupture.[10] This risk is higher in women and once the aorta is greater than 6 cm in diameter, the risk of false lumen rupture confers a 12% annual mortality.[40] Intensive lifelong surveillance is mandatory for all patients with treated or untreated aortic dissection, with many experts advocating both echocardiography and axial imaging as routine surveillance.[41] Axial imaging, preferably with MRA has been advocated at 1, 3, 6, and 12 months and yearly thereafter.[19] All patients with chronic dissection should be considered at high risk of its late sequelae. Neither open nor endovascular repair can reverse the natural history of the disease, unless the entire extent of the dissection is excluded.[42]

Conclusion

The range of options for treating patients with aortic dissection continues to expand as understanding of the condition grows. The endovascular era has heralded new hybrid procedures for the repair of acute type A dissection. The optimal management of acute type B dissection and uncomplicated chronic dissection is still medical therapy. However, endovascular technology has developed to allow feasible alternatives to traditional open surgical and medical treatments for complicated chronic dissection. Aortic dissection remains a disease with high morbidity and mortality and intensive lifelong surveillance is required for these patients.

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