DIAGNOSIS AND MANAGEMENT OF PATIENTS WITH AORTIC DISSECTION

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Cardiovascular disease is the leading cause of death in Western society and is on the rise in developing countries. Aortic diseases constitute an emerging share of the burden. New diagnostic imaging modalities, increasing life expectancy, longer exposure to elevated blood pressure, and the proliferation of modern non-invasive imaging modalities have all contributed to the growing awareness of acute and chronic aortic syndromes and pathologies.^{1–5}

Acute aortic syndrome includes aortic dissection, intramural haematoma (IMH), and symptomatic aortic ulcer. Propagation of the dissection can proceed in antegrade or retrograde fashion from the initial tear involving side branches and causing complications such as malperfusion syndromes, tamponade, or aortic valve insufficiency.⁶ Common predisposing factors in the International Registry of Aortic Dissection (IRAD) were hypertension in 72% of cases, followed by atherosclerosis in 31% and previous cardiac surgery in 18% (table 1). Analysis of the young patients with dissection (<40 years of age) revealed that younger patients were less likely to have a history of hypertension (34%) or atherosclerosis (1%), but were more likely to have Marfan syndrome, bicuspid aortic valve, and/or prior aortic surgery.⁷

DIAGNOSTIC WORK-UP

Diagnostic imaging studies in the setting of suspected aortic dissection is aimed to rapidly confirm or exclude the diagnosis, classify the extent of the dissection, and assess the emergent nature of the problem, with correct classification in distal or proximal dissection being of paramount importance (fig 1).

For confirmation of the diagnosis patients often require more than one non-invasive imaging study to characterise aortic dissection, with computed tomography (CT) used in 61% of cases, echocardiography in 33%, aortography in 4%, and magnetic resonance imaging (MRI) in only 2%.¹ Upon admission in the emergent setting transthoracic echocardiography (TTE) is useful in identifying proximal aortic dissection and thus to diagnose type A dissection in patients with shock. It is limited, however, in visualising the distal ascending, transverse, and descending aorta in a substantial number of patients. Conversely, transoesophageal echocardiography (TOE) is highly diagnostic in aortic dissection encompassing the entire thoracic aorta. Although oesophageal intubation is required, TOE can be performed at the bedside with immediate results. The diagnosis of aortic dissection is confirmed when two lumens are separated by an intimal flap.⁸ Furthermore, variants of acute aortic syndromes such as IMH and atherosclerotic penetrating ulcers can be separated with high sensitivity and specificity.²

Similarly, spiral CT scanning is accessible to most emergency departments, and provides similar (and even more extensive) information in suspected cases. In addition to TOE, CT can also assess the extent of aortic involvement and depict involvement of visceral and iliac arteries. The average sensitivity exceeds 95% with specificity ranging from 87–100%.^{9–10} The downside is the need for (nephrotoxic) iodinated contrast, radiation burden of 10–15 mSv, and the inability to assess aortic insufficiency.

MRI, although highly accurate, is of limited availability, especially on an emergency basis. Moreover, there are issues of patient inconvenience and limited applicability to patients with claustrophobia, pacemakers, aneurysm clips, or other metal devices.

Whereas retrograde aortography was the first diagnostic tool to assess patients with suspected aortic dissection between 1970 and 1980, catheter angiography is rarely performed for diagnostic purposes today, considering the better performance and safety profile of non-invasive tomographic imaging modalities. Even an incidence of 25% concomitant coronary disease fails to justify coronary angiography in the absence of data that revascularisation could improve outcome after surgery to the aorta.^{11–12}

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Table 1 Demographics and history of patients (n = 464) with acute aortic dissection

		Type A, n (%)	Type B, n (%)	p Value
Variable	n (%)	(n = 289)	(n = 175)	Type A v B
Patient history				
Marfan syndrome	22/449 (4.9)	19 (6.7)	3 (1.8)	0.02
Hypertension	326/452 (72.1)	194 (69.3)	132 (76.7)	0.08
Atherosclerosis	140/452 (31.0)	69 (24.4)	71 (42)	<0.001
Prior aortic dissection	29/453 (6.4)	11 (3.9)	18 (10.6)	0.005
Prior aortic aneurysm	73/453 (16.1)	35 (12.4)	4 (2.3)	0.006
Diabetes	23/451 (5.1)	12 (4.3)	11 (6.6)	0.29
Prior cardiac surgery	83(17.9)	46 (15.9)	37 (21.1)	0.16

Besides imaging, biomarkers for early detection of aortic dissection are recently attracting interest; a promising assay checks for circulating smooth muscle myosin heavy chain protein, a protein that is released from damaged aortic medial smooth muscle and elevated in the early hours of acute aortic dissection.¹³ Additional biochemical markers such as acute-phase reactants, C reactive protein, fibrinogen, soluble elastin fragments and D-dimer are also being studied.^{14 15}

THERAPEUTIC APPROACH

Acute dissections involving the ascending aorta are considered surgical emergencies requiring swift repair of the aortic root or reconstruction of the ascending aorta and the arch to improve prognosis. In contrast, dissections confined to the descending aorta are treated medically unless progression of dissection, intractable pain, organ malperfusion, or extra-aortic blood is demonstrated.

In the initial phase after impact the therapeutic objective is normalisation of blood pressure and lowering of the left ventricular ejection force (dP/dt), with β -blockers, to the lowest tolerable levels while ensuring adequate cerebral, coronary, and

renal perfusion. For most patients, a blood pressure between 100–120 mm Hg at a heart rate <60 beats/min is achievable. In patients intolerant to β -blockers because of asthma, bradycardia, or signs of heart failure, vasodilators and short acting calcium channel blockers are valuable options. In patients with low and even normal blood pressure at presentation, possible volume depletion from haemorrhage and/or pericardial effusion must be considered. These patients may benefit from intubation before rapid tomographic imaging for confirmatory diagnosis and swift treatment. If pericardial tamponade is diagnosed, pericardiocentesis before surgery can be harmful because it may counteract hypotonic haemostasis and eventually cause more pericardial bleeding and intractable tamponade.¹⁶

PROXIMAL (TYPE A) AORTIC DISSECTION

Acute proximal dissection (Stanford type A or DeBakey type I or II) are to be considered a surgical emergency because of the high risk of life-threatening complications (fig 1). Medical management alone has a mortality of nearly 20% by 24 h and 30% by 48 h (fig 2). Surgical treatment aims to prevent lethal complications such as aortic rupture, stroke, visceral ischaemia, cardiac tamponade, and circulatory failure. With a history of 50 years the surgical concept is to excise the intimal tear to close any entry to the false lumen, and to reconstruct the aorta with interposition of a synthetic graft with or without reimplantation of coronary arteries.¹⁷ In addition, restoration of aortic

Figure 1 Classification of distal or proximal aortic dissection. DeBakey: type I, originates in the ascending aorta, propagates at least to the aortic arch and often beyond it distally; type II, originates in and is confined to the ascending aorta; type III, originates in the descending aorta and extends distally down to the aorta or, rarely, retrograde into the aortic arch and ascending aorta. Stanford: type A, all dissections involving the ascending aorta, regardless of the site of origin; type B, all dissections not involving the ascending aorta. Adapted from Nienaber *et al.*³

Figure 2 Fourteen-day mortality in 645 patients from the International Registry of Aortic Dissection (IRAD) registry stratified by medical and surgical treatment in both type A and B aortic dissection. Adapted from Hagan *et al.*¹

Table 2 Surgical treatment of acute type A (type I and II) aortic dissection

Recommendation*	Class
Emergency surgery to avoid tamponade/aortic rupture	I
Valve-preserving surgery: tubular graft if normal-sized aortic root and no pathological changes in valve cusps	I
Replacement of aorta and aortic valve (composite graft) if ectatic proximal aorta and/or pathological changes of valve/aortic wall	I
Valve-sparing operations with aortic root remodelling for abnormal valves	IIa
Valve preservation and aortic root remodelling in Marfan patients	IIa

*All recommendations are level of evidence C.
Adapted from Nienaber and Eagle.⁴

valve competence is needed with aortic insufficiency by resuspension of the native aortic valve or valve replacement (table 2).¹⁸

With an operative mortality of 15% to 35%, surgery has a clear prognostic advantage over medical treatment.^{19–23} Adjunctive measures such as profound hypothermic circulatory arrest and selective retrograde perfusion of the head vessels have improved outcomes of proximal and arch repair.²⁴ Hypothermic circulatory arrest and retrograde perfusion have yielded mean (SD) survival rates at three and five years of 75 (5)% and 73 (6)%. Although definitive treatment of acute type A aortic dissections requires surgery, not more than 80% of patients in IRAD underwent surgery; this was because of co-morbid conditions, old age (>80 years), patient refusal, or a combination of these factors.

DISTAL (TYPE B) AORTIC DISSECTION

Patients with uncomplicated aortic dissections confined to the descending thoracic aorta (Stanford type B or DeBakey type III) are at present preferentially treated conservatively, but may be considered candidates for a reconstructive strategy such as endovascular scaffolding in the near future. Medical treatment focuses initially on haemodynamic monitoring, β -blockade, and arterial vasodilators if needed to keep systolic blood pressure <120 mm Hg, and on pain control with morphine sulfate. Once the patient is stable, oral β -blockers and other antihypertensive medications are continued under close follow-up with imaging and clinical assessment in intervals of six months. In a series of 384 patients with type B dissections, 73% were managed medically in IRAD; in-hospital mortality for these patients was

10%,²⁵ and long-term survival rate with medical treatment turned out to be 60% at five years and around 40% at 10 years.^{26, 27} Survival appeared better in patients with non-communicating distal dissections.

At present endovascular interventions for acute type B aortic dissections are generally limited to relief of life-threatening complications such as impending aortic rupture, ischaemia of limbs and organ systems, persistent or recurrent intractable pain, progression of dissection, aneurysm expansion, and uncontrolled hypertension²⁸ (fig 3). Because of extensive mortality and morbidity, classic surgery for acute type B aortic dissections has been relegated to a niche manoeuvre.²⁵ The European Society of Cardiology Task Force on Acute Aortic Dissection released recommendations for the indications for stent graft and/or fenestration in 2001 (table 3).¹⁸ Additionally, in high-risk patients not suitable for surgery because of age, co-morbid conditions, or personal preference, endovascular repair offers palliative treatment to those who otherwise would have been left to follow the natural history of the disease.

INTRAMURAL HAEMATOMA

Similar to classic type A aortic dissection, surgery is advocated in patients with type A IMH while distal or type B IMH is initially followed with medical treatment. Whereas intramural bleeding of the ascending aorta had a lower mortality with surgery (14% v 36%), patients with haemorrhage of the descending aorta had a similar mortality with medical or surgical treatment (14% v 20%).²⁹ At present, many experts recommend aortic repair for acute IMH of the ascending aorta

Table 3 Interventional treatment in aortic dissection

Recommendation*	Class
Stenting of obstructed branch origin for static obstruction of branch artery	IIa
Balloon fenestration of dissecting membrane plus stenting of aortic true lumen for dynamic obstruction	IIa
Stenting to keep fenestration open	IIa
Fenestration to provide re-entry tear for dead-end false lumen	IIa
Stenting of true lumen	
To seal entry (covered stent)	IIb
To enlarge compressed true lumen	IIa

*All recommendations are level of evidence C.
Adapted from Nienaber and Eagle.⁴

Figure 3 (A) Type B aortic dissection with imminent rupture after trauma. (B) Successful reconstruction of the thoracic aorta after stent-graft placement.

Diagnosis and management of patients with aortic dissection: key points

- ▶ Aortic syndrome comprises aortic dissection, intramural haematoma, penetrating aortic ulcer and symptomatic aneurysm
- ▶ Type A aortic dissection is considered a surgical emergency because of the high risk of life threatening complications
- ▶ Endovascular stent grafts have the potential to initiate a healing process in the setting of type B dissection by sealing the proximal entry site and depressurising the false lumen
- ▶ The concept of non-surgical reconstruction of type B dissection is being subjected to a randomised evaluation (INSTEAD)
- ▶ Long term β -blocker treatment and blood pressure control are indicated
- ▶ In the follow-up of any aortic dissection any patient with dissection should be followed by serial imaging (CT or MRI of the entire aorta)

similar to type A dissection and aggressive blood pressure lowering medication for IMH in the descending aorta similar to type B dissection.³⁰

LONG-TERM FOLLOW-UP

The 10-year actuarial survival rate of patients with an aortic dissection who leave the hospital alive ranges from 30% to 60%.^{20-23 26 27} The long-term approach is based on understanding that dissection of the aorta is the epitome of systemic aortic media degeneration or defective wall structure, with the entire aorta and its branches being predisposed to dissection, aneurysm formation, and/or aortic rupture. Subsequently, management in these patients includes life time medical treatment to minimise aortic wall stress, serial imaging to detect signs of dissection progression, redissection, or aneurysm formation, and endovascular or surgical intervention when needed.

All patients should receive lifelong antihypertensive medication including β -blockers to titrate target blood pressure to <120/80 mm Hg.³¹⁻³³ Regular imaging of the aorta should be performed one, three, and 12 months after discharge, and every 6–12 months thereafter depending on aortic size.¹⁸ The most important variables are aortic diameter, signs of aneurysm formation, and haemorrhage at the sight of surgical anastomoses or stent-graft placement. This aggressive strategy highlights that both hypertension and aortic expansion/dissection are common and not easily predicted in the first months after discharge.

In 12–30% of patients, repeated repair or reintervention is required because of extension or recurrence of dissection at the previous site of intervention, localised aneurysm formation remote from the site of repair, graft dehiscence, aortic regurgitation, or infections.³⁴ Because of the predilection for the diseased aorta to become aneurysmal at a location distant from the area of dissection, imaging of the entire aorta is advised.

CONCLUSIONS

Considering both the aging patient population in Western societies—with prolonged survival despite hypertension—and

the better diagnostic strategies available to more patients, the cardiology and cardiovascular community faces an increasing incidence of acute and chronic aortic problems that desperately need to be stratified using both early biomarkers of an inflammatory and dissecting process and functional imaging of the aortic wall. At this pivotal point in time, an elevated level of awareness in clinical cardiology and the availability of modern imaging technology should trigger an interest in diagnosing and treating the complex of acute aortic syndromes similar to previous efforts in acute coronary syndromes.

Cardiologists should improve diagnostic pathways and vascular staging in acute and chronic aortic diseases, form regional referral networks and allocation systems, and utilise uniform follow-up programmes. Moreover, precise definitions of pathology using clear semantics should be integrated into prospective registries of aortic diseases by a multidisciplinary team of physicians in an attempt to validate previous retrospective observations and to make the best use of evolving diagnostic and therapeutic strategies. Finally, cardiologists are in need of credible prognostic models that can support decisions for individual patient care independent of investigators, at different times, and in worldwide locations.

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