Acute Aortic Syndromes

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"…the pericardium was found distended with a quantity of coagulated blood, nearly a pint…; the whole heart was so compressed as to prevent any blood contained in the veins from being forced into the auricles; therefore the ventricles were found absolutely void of blood…; and in the trunk of the aorta we found a transverse fissure on its inner side, about an inch and a half long, through which some blood had recently passed under its external coat and formed an elevated ecchymosis."

Frank Nicholls (physician) on the death of King George II of Great Britain - 1760

Definition

Acute **A**ortic **S**yndrome refers to a characteristic clinical presentation caused by life threatening acute aortic pathologies including **A**cute **A**ortic **D**issection (**AAD**), **I**ntra**M**ural **H**aematoma (**IMH**) and **P**enetrating **A**ortic **U**lcers (PAU).

Acute Aortic Dissection (AAD)

Aortic Dissection is the commonest aortic emergency (70% of AAS)[1] and occurs when a tear develops in the aortic intima (inner aortic wall layer) allowing blood to pass between the aortic wall layers creating a false tract between the intima and the media (middle layer) or adventitia (outer layer). The dissection can progress in a retrograde or an antegrade fashion and cause complications including aortic rupture, pericardial tamponade, aortic valve regurgitation, ACS, acute neurological syndrome, renal failure and bowel or limb ischaemia[2,3,4].

Intramural Haematoma (IMH)

IMH originates from a ruptured vasa vasorum in medial wall layers. The bleeding into the medial layer forms the haematoma. This may lead to infarction of the intima and a tear causing aortic dissection[5,6]. IMH represents up to 20% of AAS[5,7]. It may progress, dissect, regress or even resorb[7].

Penetrating Atherosclerotic Ulcers

PAUs are deep ulceration of atherosclerotic plaques penetrating the intima. The ulcers can be single or multiple and can progress to IMH, dissection, aneurysm or perforation. PAUs are often found in the descending aorta.

Epidemiology

The incidence of aortic dissection is 2.6 to 3.5 cases per 100,000 person/year[8]. **IRAD** (**I**nternational **R**egistry of **A**cute **A**ortic **D**issection) report indicates that the mean age of presentation is 63 years and males are twice affected as females. The most common risk factor is hypertension 75%, followed by atherosclerosis, found in 31%[1]. Table 1 shows common risk factors.

Table 1: Risk Factors for Aortic Dissection

Long-standing hypertension
Male
Advanced age
Prior cardiac surgery
Known aortic aneurysm or prior dissection
Cardiac catheterization/surgery
Connective tissue disorders - Marfan's syndrome - Ehlers−Danlos syndrome - Bicuspid aortic valve - Coarctation of the aorta - Hereditary thoracic aortic disease
Vascular inflammation
Deceleration injury
Cocaine
Peripartum

Classification

Accurate classification helps in deciding medical versus surgical treatment. There are two classification systems for aortic dissection. Stanford classification (Table 2) can be used for AAD, IMH and PAU. DeBakey classification (Table 3) is another classification used in aortic dissection.

Table 2	Stanford Classification
Stanford A	Involves the ascending aorta and may or may not involves the descending aorta
Stanford B	Involves the descending aorta and spares the ascending aorta

Table 3	DeBakey Classification
DeBakey type I	Originates in ascending aorta and extends at least to aortic arch
DeBakey type II	Confined to ascending aorta
Debakey type III	Originates in descending aorta and extends distally, rarely to ascending aorta

Clinical Presentation

The most common symptom is sudden onset severe pain most commonly described as sharp or stabbing[9,10]. It can occur in the chest, back or abdomen depending on the site of the disease. It can radiate to the neck, throat or jaw indicating ascending aorta involvement. Back pain and abdominal pain indicate descending aorta involvement[11]. The dissection can progress to the aortic valve causing aortic valve regurgitation, pericardium causing tamponade, carotid branches causing stroke, renal arteries causing renal failure or iliac branches causing limb ischemia. Blood loss can lead to syncope. Other signs include high blood pressure and pulse deficit. If the dissection involves the coronary arteries patient may present with an acute myocardial infarction.

Investigations

The use of biomarkers to diagnose AAS is very attractive but it is currently still undergoing active research and development. DDimer has a negative predicting value of 100% but specificity is very low[12]. Serum smooth muscle myosin heavy-chain protein levels rise initially, but rapidly clear within 24 hours[13].

ECG

It is important to rule out other causes. However it is frequently abnormal (70%). Common findings include nonspecific ST- T wave changes (42%), ischaemic changes (15%), acute MI in (5%) and LVH[11].

Imaging

The aims of imaging are:

1- Confirm the Diagnosis.
2- Locate the intimal tear if present.
3- Assess the extent of the dissection.
4- Classification.
5- Look for complications.

An IRAD report showed that two thirds of patients undergo multiple imaging and 98% of patients undergo one of TTE, CCT/MDCT, MRI and Aortography[11].

CXR

85% of patients have widened mediastinum. Other features include pleural effusion and cardiomegaly. However, 10 -15% of patients have a normal CXR[14].

Aortography

Previously used for diagnosis but is now over placed by other imaging techniques.

TTE

It is useful in detecting proximal dissection, screening patients with shock and detecting aortic regurgitation, pericardial tamponade, and heart failure but has limited value in the evaluation of part of the ascending aorta, aortic arch and abdominal aorta.

TOE

The second most commonly used (33% in IRAD report). Its sensitivity is 90% for type A but only 80% for type B[15]. It is a relatively quick modality and doesn't require patient transport. The proximity of the oesophagus to the aorta allows clear vision of the thoracic aorta. The limitations include a blind area due to the air in the trachea covering the aortic arch, inability to assess the abdominal aorta, sedation, retching and gagging can cause bradycardia and elevate the blood pressure, can not be used in patients with oesophageal disease and the need for a trained cardiologist to perform the procedure.

CT, MDCT or CTA

Most frequently used modality and has a sensitivity is 93% for both Type A and Type B Aortic Dissection[16]. It provides anatomic information regarding the aorta and surrounding structures as well as identifying aortic rupture. Nearly two thirds of patients have a CT scan as a first line imaging modality. Disadvantages include need for transport, its inability to evaluate aortic valve and coronary arteries, motion artefact due to the heart motion and contrast induced nephropathy.

MRI/MRA

Used in <5 % of patients in IRAD[13] but is highly sensitive for both Type A and B. It is useful in detecting PAU and IMH and visualizes the main aortic branches and surrounding structures. It is often used for follow up and has the advantage of no radiation. However, its limitations include availability in emergency settings and its contraindication in patients who have some implantable devices, clips, metallic valves and claustrophobia.

Treatment

Initial treatment

The patients are best treated in a highly monitored unit unless immediately transferred to an emergency theatre. The aim is to stabilize the patient haemodynamically and control the blood pressure. The blood pressure should be intensely managed and the heart rate should be controlled while signs of shock, arrhythmias and other complications are observed closely. Type A aortic disease is treated surgically while the gold standard for treating uncomplicated Type B acute aortic syndromes is medical treatment.

Medical Treatment

Medical treatment is the preferred treatment option for patients with stable descending aorta disease (Type B). This is due to both the lower mortality rate in patients with stable disease compared to unstable patients, and the high operative mortality rate. The aim is to control the systolic blood pressure to a target of 100 -120 mmHg and to slow the heart rate closest to 60 bpm reducing the stress on the aortic wall. Intravenous β-blockers are the first line therapy and Labetalol is commonly used due to the combined α and β blocking characteristics. It causes vasodilatation and reduces the left ventricular ejection force. Esmolol has a short half life of 0.13 hours and can be used for testing β-blockers tolerance or in patients who are awaiting operations. Often dual therapy is needed and intravenous Sodium Nitroprusside has been frequently used due to its vasodilator activity. However, if used alone it can cause sudden increase in left ventricular ejection force and worsen the aortic disease. Therefore, it is commonly used in combination with β-blockers. If β-blockers are not tolerated calcium channel blockers are used as a second line. Verapamil and Diltiazem have vasodilator effects and reduce the heart rate. Pain secondary to the acute aortic syndrome can cause hypotension and tachycardia therefore adequate analgesia is needed and intravenous morphine is commonly used.

Surgical Treatment

Type A Acute Aortic Syndromes

Type A acute aortic dissection has a mortality rate of 1 -2% per hour during the first 24 hours[11]. If treated with medical management alone the mortality rate is 20% at 24 h, 30% at 48 h and 50% at 1 month. Surgical treatment is the goal standard for type A AAS and the aim is to

prevent or treat complications. Open surgery involves excision of the intimal tear and replacement of the ascending aorta (Figure 6). Other procedures include aortic arch replacement, aortic valve replacement or resuspension and coronary artery bypass. Operative mortality rate vary between 10-35% which is lower than the 50% rate if treated medical alone[17,18].

Type B Acute Aortic Syndromes

The mortality rate for patients treated with medical therapy alone is 10%[19]. Surgical treatment is reserved to patients developing complications, refractory pain, refractory hypertension or progression of dissection. Endovascular treatment is emerging as superior to open surgery however there is not enough evidence to support favourable long term outcomes. IRAD report shows that in acute type B aortic dissections in-hospital mortality rate is 7% in patients who had endovascular repair and 32% in patients who had open surgery[11].

Endovascular treatment

It is more commonly used for complicated type B aortic dissection, IMH and PAU. For type A aortic dissection it is reserved to patients who refuse open surgery or who are not fit for open surgery. The procedure involves fenestration and/or stent placement which allows decompression of the false lumen, re-expansion of the true lumen and facilitate closure of the intimal tear which in turn induces thrombosis in the false lumen.

Follow up

Patients with AAS have a 10 year mortality rate ranging between 30 to 60%[19,20]. They have a life long risk of further complications. Factors contributing to late complications include hypertension, age, aorta size, genetically inherited connective tissue disorders and presence of a patent false lumen[21,22]. Hypertension should be controlled and β-blockers are commonly used. Angiotensin converting enzyme inhibitors, calcium channel blockers and diuretics are used but there is no evidence to support secondary prevention. Follow up imaging is recommended at 1, 3, 6, 9 and 12 months followed by annual imaging[23]. Modalities commonly used are MRI and CT scan.

Outlook

- The use of biomarkers in diagnosing acute aortic syndromes is attractive but still undergoing active research.
- Early diagnosis and management of acute aortic syndromes is important in reducing mortality and currently there is lack of clinical pathways for managing AAS.
- Advances in therapeutic procedures continue to develop and the role of endovascular treatment is increasing.

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