AORTIC DISSECTION

Field Guide for Primary Care

Ensuring that everyone with aortic dissection has their opportunity to live a better quality of life

Primary Care Cardiovascular Society

Driving primary care to deliver the best in cardiovascular health

THE AORTIC DISSECTION CHARITABLE TRUST

www.tadct.org

Contents

Foreword

The Aortic Dissection Charitable Trust is the UK and Ireland charity dedicated to aortic dissection. This document, produced in collaboration with the Primary Care Cardiovascular Society (PCCS), supports the aims of the Trust to bring consistency of treatment across the whole patient pathway through education, policy change and fostering research.

The PCCS is delighted to be working with the Trust in producing this field guide to aortic dissection, to ensure that everyone with aortic dissection has their opportunity to live a better quality of life. Dr Victoria McKay, Consultant Clinical Geneticist, Liverpool Heart and Chest Hospital NHS Foundation Trust and Dr Bejal Pandya, Consultant Cardiologist, Barts Health NHS Foundation Trust both made valuable contributions to this guide for which we are grateful. Both organisations look forward to working together on future projects and with the wider health service. And in doing so, we're confident we can improve the lives of people living with aortic dissection across the UK and Ireland.

Dr Jim Moore
President
Primary Care Cardiovascular Society

Graham Cooper MD FRCS (CTh)
Trustee
The Aortic Dissection Charitable Trust

Pathology

Aortic dissection occurs when a tear in the lining (intima) of the aorta allows blood to enter the aortic wall. This creates a separation in the layers (media) of the aortic wall, which creates a false lumen through which blood can flow, in addition to the natural flow through the true lumen.

The length of the aorta through which the false lumen extends is variable, but typically the entire aorta beyond the entry tear is involved.

Acute events affecting the thoracic aorta are termed acute aortic syndromes, with dissection accounting for 80 to 90% of events, and intra-mural haematoma accounting for approximately 10% of cases. Intra-mural haematoma is thought to occur due to rupture of blood vessels in the aortic wall (vaso vasorum), with resultant clot formation in the wall. In the acute phase, intra-mural haematoma is essentially treated in the same way as aortic dissection.

In the acute phase, aortic dissection is a dynamic process, which continues until a stable balance is achieved between the flow and pressure in the true and false lumina. The establishment of this balance usually involves the development of further intimal tears beyond the initial entry tear, known as re-entry tears. These allow blood to re-enter the true lumen from the false lumen, which will equalise pressure between the two. Transient or prolonged interruption of blood flow to aortic branches may cause visceral, limb, cerebral or cardiac ischaemia, which is termed malperfusion.

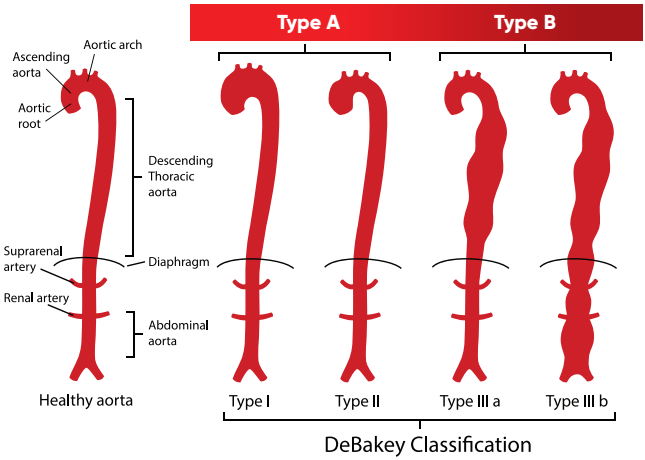
In the chronic phase, blood flow through the true and false lumina is stable. The principle ongoing risk in this situation is the expansion of the false lumen to a size where there is a risk of rupture.

Classification

The classification of aortic dissection is determined by the location of the entry tear and the extent of the aorta involvement.

The Stanford classification describes type A dissection as where the entry tear is within the ascending aorta, and type B dissection as where it lies in the descending thoracic aorta.

In the DeBakey classification, in type I and II the entry tear is in the ascending aorta, in type I the dissection extends into the descending aorta, and in type II it is confined to the ascending aorta, whereas type III involves only the descending aorta. In terms of the various classifications, 70% of dissections are Stanford type A.



70% of dissections are Stanford type A

Incidence and Prevalence

There are many papers describing incidences of aortic dissections, with ranges between 3 and 12 per 100,000 population.

The strongest data is from the Oxford Vascular Study, which gives an incidence of 6 per 100,000 new aortic dissections per year. The incidence increases with age, and this represents about 4000 new cases per year in the UK, of which almost 3000 are type A.

Prevalence is much less well described, with data from Taiwan giving a prevalence of 20 to 30 cases per 100,000 population.

The incidence and prevalence of aortic dissection are both increasing, largely due to ageing of the population. The Oxford Vascular Study estimates that the incidence of aortic dissection will double by 2050.

Aetiology

Hypertension, especially untreated hypertension, is responsible for around 70% of cases. There are a range of aetiologies for the remaining cases:

- Genetically triggered thoracic aortic disease:
  - Marfan syndrome
  - Bicuspid aortic valve
  - Loeys-Dietz syndrome
  - Familial thoracic aortic dissection
    There are a range of gene changes associated with familial thoracic aortic dissection
  - Vascular Ehlers-Danlos syndrome
  - Coarctation of the aorta
  - Turner syndrome
- Trauma
- Previous cardiac surgery
- Cocaine use
- Inflammatory/infectious diseases
  - Giant cell arteritis
  - Takayasu arteritis
  - Bechet disease
  - Aortitis
- Pregnancy

Presentation

Presentation is similar for both type A and B dissection, and also intra-mural haematoma.

The onset of acute aortic dissection is characterised by the sudden onset of severe chest, back, neck or abdominal pain, which often radiates between these sites. The pain is maximal at onset and may settle quickly. Symptoms due to malperfusion occur in around 20% of patients, these are often transient and may confound making the diagnosis. In addition, acute type A dissection may cause new aortic regurgitation.

Diagnosis requires a CT scan and once made requires emergency specialist treatment.

Occasionally, dissection is not identified in the acute phase and is identified incidentally on a CT scan undertaken for other reasons.

In the chronic phase, provided adequate surveillance is undertaken, expansion of the false lumen is very unlikely to reach a size that will give rise to symptoms. If symptoms did arise, they would be those associated with any large thoracic aortic aneurysm; back pain, hoarseness, dysphagia, and shortness of breath are typical. Very rarely patients may suffer a second acute dissection within an already dissected aorta.

The onset of acute aortic dissection is characterised by the sudden onset of severe chest, back, neck or abdominal pain

Joanne's Story

Joanne was 39 when she had a type B aortic dissection. She had open-heart surgery for valve repair with an aortic root replacement (David procedure) at 35 because of Marfan Syndrome.

"On the morning of 31st October 2012, I turned over in bed, I had an awful pain in my chest which was followed by a severe tearing feeling, going from the top left of my body, down to the bottom right and left me feeling clammy and nauseous. I knew I was very unwell and asked my husband to call an ambulance. I was taken to my local A&E department, where I was monitored and given an ECG and X-ray. After having substantial pain killers, I was discharged after about two hours, with the advice 'take pain killers every four hours'.

I spent the next days in considerable pain, taking regular painkillers, but still felt very unwell. I was persuaded to go to see my GP two days after the original event.

My GP listened to my story and after a short wait, arranged for me to be admitted to a day ward at a different local hospital, he had immediately suspected there was something seriously wrong.

I was admitted to the ward and given, almost immediately, a CT scan. This showed up the considerable type B dissection."

Treatment

For acute type A dissection, the treatment is emergency cardiac surgery. At a minimum this will involve replacement of the ascending aorta, but in more complicated cases may involve replacement of the sinuses of the Valsalva with reimplantation of the coronary arteries, and possibly replacement of the aortic valve and/or aortic arch replacement.

Type B dissection can usually be treated medically with blood pressure control. However, in around 15% of patients the dissection is complicated by persistent pain, malperfusion, rupture, and/or uncontrollable hypertension and intervention is indicated. This involves the placement of a covered endovascular stent, which us used to cover the entry tear in the descending aorta. Occasionally, depending on the precise location of the tear, a bypass of the left subclavian artery may be required.

Survival

About a 50% of patients with an acute type A dissection die before reaching hospital, usually due to rupture of the aorta into the pericardium and the consequent cardiac tamponade.
For those undergoing surgery, operative survival is about 80%.

For type B dissection, about 90% of patients survive to reach hospital. 10-year survival is about 50% at 10 years, but with significant variation due to the wide age range that aortic dissection affects.

The risk of further intervention on the dissected aorta is about 10% at 5 years and 40% at 10 years.

Follow Up

Survivors of aortic dissection should be followed up in a specialist clinic. They will require annual imaging of their aorta, by either CT or MRI.

This is to detect progressive dilatation of the dissected aorta. The threshold for intervention is, in general, a diameter of 6.0cm but this may be less in patients with connective tissue disorders, or more in those of advanced age or with significant comorbidities.

About 40% of patients who have suffered an aortic dissection have some manifestation of post-traumatic stress disorder.

Exercise

Due to the blood pressure response, vigorous dynamic (aerobic), and static (isometric) exercise should be avoided, as elevated blood pressure places shear stress on the aortic wall causing expansion.

In syndromic aortopathies, the systolic blood pressure should be lowered to 100mmHg at rest and 120mHg on exertion. Dynamic exercise (aerobic) is safer post dissection as it increases heart rate rather than blood pressure to increase cardiac output. Static (isometric) exercises are contraindicated as they can cause a vast increase in blood pressure. Heavy weight training should be completely avoided. In addition, resistance training results in increased intra-abdominal pressure and a Valsalva manoeuvre, which, when released also causes a surge in blood pressure making it inadvisable. The European Society of Cardiology recommend that no more than 5 METS are exercised post aortic dissection, this is equivalent to enthusiastic dancing. **Although, moderate aerobic exercise should be encouraged.**

Sex has a moderate effect on heart rate and can raise systolic BP up to 40mmHg at orgasm, which normalises within 2 minutes.

Treatment
in the Chronic Phase

Good blood pressure control is vital, and the target blood pressure should be

< 130/80 mmHg

A beta blocker is the first choice of anti-hypertensive medication.

ARTERIAL BLOOD PRESSURE RESPONSE TO HEAVY RESISTANCE EXERCISE		
	Dynamic	**Static**
Peripheral Vascular Resistance	Decrease	Increase due to vasoconstriction
SBP	140-180mmHg	Up to 480mmHg
Mean BP	Modest Increase	Large increase (DBP350)
After Exercise Effect	Return to Normal	Surge in BP after Valsalva
Aorta in Athletes	Enlarged in Elite	Enlarged

Ref: MacDougall JD, Tuxen D, Sale DG, et al. J Appl Physiol 1985. Palatini et al, Journal of Hypertension 1989.

WHAT EXERCISE SHOULD BE AVOIDED?			
BOXING	**RUGBY**	**DEEP SEA DIVING**	**ROWING**
DISTANCE RUNNING	**SKY-DIVING**	**HANG GLIDING**	**MOUNTAINEERING**
TRAMPOLINING	**HIGH DIVING**	**WEIGHTLIFTING**	**KARATE**
JUDO	**WRESTLING**	**SQUASH**	**BASKETBALL**

Genetic Testing

Diagnostic genetic testing should be offered to any patient with:

- Thoracic aortic aneurysm (a diameter greater than 3.8cm) or dissection under the age of 50 years.
- Thoracic aortic aneurysm or dissection under the age of 60 years with no cardiovascular risk factors.
- Thoracic aortic aneurysm or dissection under the age of 60 years with a family history of thoracic aortic aneurysm or dissection in a first-degree relative.
- Any syndromic features suggestive of Marfan syndrome, Loeys-Dietz syndrome, vascular Ehlers Danlos syndrome or related conditions.
- Any deceased individual meeting any of the above criteria where DNA for testing is available.

Patients meeting the above criteria should be referred to a Regional Clinical Genetics, a Genomics Service, or an Inherited Cardiovascular Conditions Service.

Where a patient chooses to go ahead with a diagnostic genetic test and a conclusive genetic change causative of the aortopathy is identified, first-degree relatives will be invited to come forward for a predictive genetic test. Predictive genetic testing allows relatives to determine whether they are also at-risk of aortic disease and if a clinical follow up is required.

Where a patient chooses to go ahead with a diagnostic genetic test, and no genetic changes or an inconclusive genetic change are identified, family clinical surveillance will usually still be recommended.

Clinical Surveillance

First degree relatives of those with a bicuspid aortic valve should undergo echocardiography to exclude this.

In families where a person meets the genetic testing criteria described above, but where a genetic cause has not been identified, clinical surveillance and follow-up should be offered to all first-degree relatives. This includes parents, children, and siblings (biological brothers and sisters), who will usually be offered a clinical follow up and imaging with echocardiography and/or cross-sectional imaging in the form of CT or MRI.

Some treating centres have a cardiology-led aortopathy surveillance clinic, who will arrange ongoing follow up.

Acknowledgements

The Aortic Dissection Charitable Trust is very grateful to:

Dr Victoria McKay Consultant Clinical Geneticist, Liverpool Heart and Chest Hospital NHS Foundation Trust

Dr Bejal Pandya Consultant Cardiologist, Barts Health NHS Foundation Trust

for their valuable contribution to this document.

The Primary Care Cardiovascular Society

The Primary Care Cardiovascular Society (PCCS) is a UK-based multi-disciplinary organisation reflecting the wide range of primary care healthcare practitioners throughout the UK involved in delivering cardiovascular patient care and improving cardiovascular health.

We are committed to promoting best practice in primary care cardiovascular healthcare through high quality education, training and service development.

We support the development and education of primary healthcare practitioners in cardiovascular medicine and have a role in facilitating primary care cardiovascular research. Members of the PCCS Council and the wider society membership are involved in key organisations and working groups at a local, regional and national level including NICE.

Membership is free for practising UK healthcare practitioners and gives access to our, members only, CVD Academy offering educational opportunities in the form of online workshops focused around key CVD areas, with downloadable CPD certificates, and resources (including latest guidance and relevant publications).

Primary Care Cardiovascular Society

Driving primary care to deliver the best in cardiovascular health

www.pccsuk.org

The Aortic Dissection Charitable Trust

The Aortic Dissection Charitable Trust aims to improve the diagnosis of aortic dissection and bring consistency of treatment across the whole patient pathway.

We accomplish this through:

- Increased access to education for medical professionals and patients in the UK & Ireland
- Working with those responsible for healthcare policy in the UK & Ireland to ensure that there is consistency in the provision of diagnosis for acute aortic dissection, specialised follow-up for survivors and access to clinical genetics for relatives
- Promoting funding for medical research into the detection, prevention, treatment and cure for aortic dissection

THE
AORTIC DISSECTION
CHARITABLE TRUST

www.tadct.org

This guide is supported by the:

Primary Care Cardiovascular Society

Driving primary care to deliver the best in cardiovascular health

TAD CT

@AorticDissectCT

www.tadct.org

UK Charity Number 1191420