

**Acute Aortic Syndrome Pathway:** *recommendations for diagnosis, early management, referral and transfer within the Merseyside & Cheshire STP, North Wales and Isle of Man.*

**Short title:** Acute Aortic Pathway

**Authored by:** Mark Field on behalf of *Liverpool Acute Network for Thoracic Aortic Services (LANTAS)*

**Commissioned by:** Jane Tomkinson CEO LHCH and Chair CVD Merseyside and Cheshire STP

**Local referral patterns:**

-Cheshire and Merseyside (STP)

-Isle of Man

-North Wales

**National Referral patterns:**

Nationwide

Approved by Aortic Dissection Awareness UK

## **Liverpool Acute Network for Thoracic Aortic Disease**

### **(LANTAS)**

#### **The Team**

##### *Surgeons and Interventional Radiologists*

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**Aortic Network Coordinator:** Jamie Doolan LHCH

**Second Wednesday Monthly MDT:** 0815 Radiology Seminar Room, LHCH

## **Background**

Acute Aortic Syndrome (AAS) is a collection of acute aortic pathologies that includes acute Type A aortic dissection, acute Type B aortic dissection, intramural haematoma, penetrating atherosclerotic ulcers and blunt trauma related injury. All have different risks and require different management pathways. Acute Type A pathologies involve the ascending aorta and typically require immediate surgery. Type B pathologies involve a variable extent of the thoracoabdominal aorta with uncomplicated presentations managed medically and complicated presentations with leaks or malperfusions managed urgently. Often acute on chronic pathologies represent more complex scenarios requiring bespoke patient specific approaches.

Within the Merseyside and Cheshire STP lays the Liverpool Heart and Chest Hospital with a specialised Thoracic Aortic Aneurysm Service and “LiVES” (Liverpool Vascular and Endovascular Services) comprising a network of vascular surgeons and interventional radiologists localised between Royal Liverpool University Hospital, Aintree University Hospital, Southport Hospital and Ormskirk and St Helens and Knowsley Trust.

In addition to the Merseyside and Cheshire STP area our catchment area includes North Wales and Isle of Man, however patients are referred from neighbouring catchment areas in Lancashire (Blackpool), Manchester (MRI and Wythenshaw) and Staffordshire (Stoke on Trent) and also more distant regions and in particular Scotland, North East and Midlands. This document sets out our recommendations for diagnosis, early management, referral and transfer of acute aortic pathologies into LANTAS.

<http://www.lhch.nhs.uk/our-services/aortic-aneurysm-surgery/information-for-referring-doctors/>

## **Acute Aortic Pathologies**

### ***1) Type A Acute Aortic Syndrome (AAS)***

- a. Acute Type A aortic dissection
- b. Acute Type A Intramural Haematoma (IMH)

### ***2) Type B Acute Aortic Syndrome***

- a. Uncomplicated dissection
- b. Complicated dissection
- c. Intramural Haematoma
- d. Penetrating Aortic Ulcer (PAU)

### ***3) Acute on chronic thoracoabdominal aortic disease***

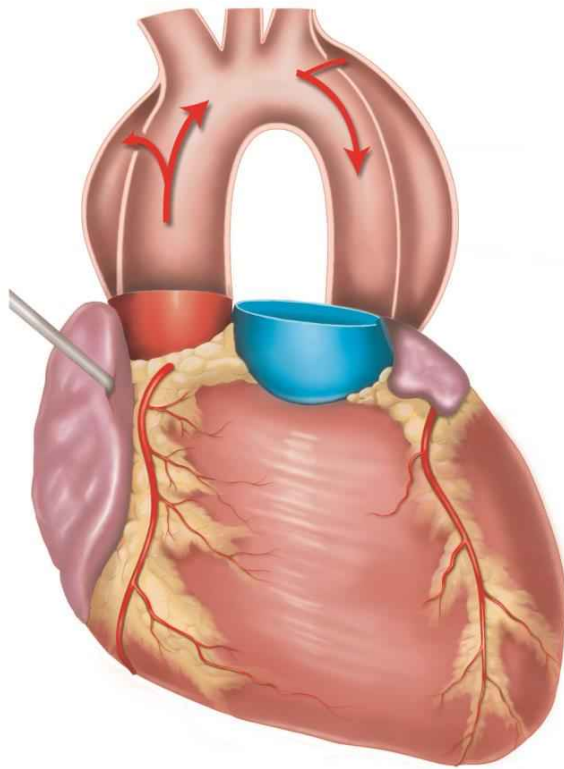
### ***4) Other***

- a. Mycotic aetiologies
- b. Trauma

## **Acute Type A aortic dissection or IMH**

### **Pathology**

Acute Type A aortic dissection is a splitting of the tunica media resulting in a true lumen and a false lumen.



**Figure:** *Acute Type A involves the ascending aorta while acute Type B does not.*

The natural history following the index event is poor with a 1% per hour mortality over the first 48 hours. Patients die of rupture and tamponade, aortic insufficiency and heart failure as well as malperfusion syndromes of the coronaries, cerebral circulation, viscera and limbs. Crucial to a successful outcome is early diagnosis, immediate medical management and transfer to the nearest specialist centre for surgery. Intra-mural haematoma is a bruising of the aorta due to rupture of the vasa vasorum and has been shown to carry the same risk profile as formal aortic dissection and requiring surgery. While surgical and anaesthetic techniques have improved and with them outcomes, there remains a stubbornly high mortality even in specialist centres. The reasons for this are three fold:

- 1) Delayed diagnosis and end-stage disease on arriving in theatre
- 2) Sub-optimal initial medical management
- 3) Delays in transfer between hospital and into the operating room.

This document is intended to offer guidelines to improve these three remaining challenges.

## **Presentation**

Patient may present with classical chest pain striking from front to back between the scapula blades. However, they may present in a variety of ways including atypical chest, stroke, heart failure, acute abdomen and limb ischaemia, depending on the extent of dissection, degree of end-organ involvement and degree of malperfusion. As aortic dissection is a rare disease and represents only a very small proportion of patients presenting with chest pain, the key to making the diagnosis is to always “think aorta”.



## **Diagnosis**

The rarity of acute Type A is the Achilles heel of the disease. Doctors in A&E will rarely if ever see such a patient. Campaigns encourage doctors (<http://www.aorticdissectionawareness.com/aortic-dissection/>) to think aortic dissection however by far the biggest reasons for chest pain are myocardial infarction, pulmonary embolism and non-specific musculoskeletal aetiologies. Standard investigations for chest pain will include: bloods (incl. cardiac enzymes and D-dimers), ECG and CXR with more advanced imaging including echocardiography and CT scans. Standard investigations together with a thorough history and examination should lead the doctor to at least suspect aortic dissection and order specialist investigations particularly CT. Echo is ideal but not mandated and should not delay the management of the patient. Evidence for malperfusion should be assessed once the diagnosis is made.

## **Early management**

Patients need immediate intravenous access, analgesia and blood pressure control. Ideally all patients should have:

- 1) IV access
- 2) Labetalol infusion
- 3) Catheter
- 4) Arterial line

This is important to reduce risks of rupture or extension of the dissection as well as allow safe transfer.

## **Referral**

All acute Type A pathologies should be referred to LHCH. At the Liverpool Heart and Chest Hospital there are a numbers of ways to refer in however the fastest pathway is to ring switchboard (0151 2281616) and ask to be put through to the Aortic Fellow. The Aortic Fellow will take the details and request transfer of imaging CT scan. If the Aortic Fellow cannot be contacted then the Senior Surgical SpR should be contacted by switchboard. Failing that, or if there is a desire to speak directly to the consultant on-call aortic surgeon, switchboard

will put you through to one of Mr Kuduvalli, Mr Field, Miss Harrington or Mr Nawaytou.

Ideally we would prefer an ECG gated CT entire aorta. Gated CT scans may not always be possible and if there is any question about non-gated motion artefact in the images of the root which give the appearance of a “pseudo-dissection” the patient may require transfer for a gated scan. On occasions the diagnosis may be made on a CT PA or on a CT scan which doesn’t cover the entire aorta. In these circumstances it will be preferable to transfer the patient to our hospital for further specialist images rather than delaying transfer. On occasions there may be issues transferring the images on PACs via the Link. Should this be the case, transfer should be based on the report and image transfer should not delay patient transfer. We do need to see the images before taking the patient to theatre and therefore as a last resort the patient should be sent along with a CD containing the appropriate images along with the password for access. If the patient arrives with no access to images a repeat CT will be required which will delay surgery.

## **Transfers**

Once accepted by the surgeon, there will need to be a discussion with our on-call Intensivist regarding an ITU bed. The Matron in our ITU will contact the nurses in the referring hospital to arrange a blue light transfer. The patient must not be put in an ambulance and transferred until our nurses confirm a bed is available, or a plan to admit to theatre.

Ideally the patient should be transferred with a medical escort with monitoring (IV access, arterial line and catheter) as well as strict BP control with Labetalol. There is no room for a so-called “scoop and run” approach. A safe and stable transfer is essential.

## **Destination within LHCH**

Patient will normally be transferred into the Critical Care Area which may be: 1) Intensive Care, 2) Post Operative Care Unit (POCCU) , or 3) Coronary care Unit. For some selected patients the destination will be directly through the Critical Care Area into the Theatre Suit. These patients will be those identified at very high risk:

- 1) Peri-arrest
- 2) Obvious tamponade
- 3) Evidence of myocardial infarction
- 4) Evidence of general low cardiac output and/or malperfusion  
(lactate >4)
- 5) Evidence of limb ischaemia

These features should be identified to the surgeon taking the referral. More stable pain free patients with no compromise will be assessed on ITU first. This is particularly the case for stable patients with a delayed (over days) diagnosis or patients who have sustained a stroke whom in which a more measured, timely intervention may be considered.

## **Standard Setting**

### **Key Quality Markers**

#### **1) Door to skin within 6 hours**

#### **2) Diagnosis**

- a. Diagnosis within 4 hours of arrival

#### **3) Early management**

- a. All the following:
  - i. Labetalol infusion
  - ii. Arterial line
  - iii. Catheter
  - iv. IV access

#### **4) Transfers**

- a. Transfer to centre and theatre within 2 hours

### **Feedback to referring centres in the network**

Feedback to referring A&E will be on an annual basis to the head of Department and benchmarked against our KQI.

### **MDT**

All patients will be discussed at the next Aortic MDT for consideration of distal intervention in the sub-acute phase.

### **Follow-up**

All patients will be offered life long follow-up and as appropriate, referral to clinical genetics.

## ***2) Type B Aortic dissection or IMH***

- a. Uncomplicated dissection
- b. Complicated dissection

### **Pathology**

Acute Type B aortic dissection, like acute Type A, is a splitting of the tunica media however not involving the ascending aorta. The risk profile is different to acute Type A aortic dissection without the risk of cardiac tamponade, aortic valve insufficiency or myocardial infarction. The default management is medical and associated mortality is around 10-15% for uncomplicated disease. If there is malperfusion of spinal cord, viscera vessels, renal vessels or limbs or evidence of contained rupture then intervention may be required.

The challenge with acute Type B aortic dissection is not just diagnosis, early management and transfer as with acute Type A, but managing the heterogeneity of specialities involved and stewarding the patient into a regional single point of contact (SPOC) and MDT discussion. Those specialties involved in diagnosis and care often



include: Accident and Emergency, General Medicine, General Surgery, Cardiology, Intensive Care, Vascular Surgery and Cardiac Surgery. The pathway to specialist care is equally diverse in the region with most hospitals caring for uncomplicated acute Type B aortic syndromes under the advice of LANTAS and complicated presentations transferred into RLUH or LHCH.

### **Presentation and Diagnosis**

Patients present in a vast variety of ways but principally chest pain and as such the difficulty like for acute Type A aortic dissection is in diagnosis and identifying them from myocardial infarction, pulmonary embolism and non-specific chest pain. Patients may present in other complex ways relating to visceral malperfusion and limb ischaemia. History and examination and a differential diagnosis of aortic dissection are key. Standard investigations will include: ECG, cardiac enzymes, D-dimers, CXR however the diagnosis is made on CT scan. A contrast CT of the entire aorta is required.

## **Early Management**

The pathway for patients is dependent on the extent of the dissection and the presence or not of malperfusion.

### **1) Uncomplicated**

Patients diagnosed with uncomplicated acute Type B aortic dissection or IMH will be managed medically and with serial imaging typically in their presenting hospital under the advice of LANTAS. Patients should be admitted to a Critical Care Area and require:

- 1) IV access and analgesia**
- 2) Arterial BP monitoring and Labetalol infusion**
- 3) Urinary catheter**

The aim of management is to steward the disease into a chronic state to enable monitoring and follow-up on an out patient bases and possible elective aneurysmal surgery or TEVAR. Patients should be monitored for signs of progression of the AAS whether that be malperfusion or leak.

Serial CT scanning is the only way to accurately monitor the disease in the acute phase and we recommend that after the initial diagnostic CT scan further scans are performed at 48 hours and 5 days. This may be modified of course if there is on-going pain or a suspicion of disease progression.

During this period the patient should be established on oral antihypertensive therapy and will typically include B-blockers, calcium channel blockers, ACE inhibitors and alpha blockers. Providing the patients BP is well controlled, pain has settled and CT scans are satisfactory, they may be discharged home to be seen in clinic at LHCH at 4 weeks with a CT on arrival. A formal letter of referral should be sent on discharge. The patient will be discussed at the next Aortic MDT at LHCH and considered for TEVAR in the sub-acute phase (2-12 weeks).

Contact: Initial contact should be either LHCH or RLUH. At LHCH ask Switchboard for the Aortic Fellow on-call. If this fails other points of contact are the Cardiac Surgical SpR on-call, Hospital Coordinators or Aortic ANP Coordinator (Jammie Doolan).

Alternatively the consultant aortic surgeon on-call may be contacted directly. At RLUH there is a Vascular SpR on-call or the Vascular Surgeon may be contacted directly.

## 2) Complicated

Whether the initial diagnosis is of a complicated acute Type B aortic syndrome or the disease progresses under follow-up, transfer is warranted to LHCH or RLUH.

Complicated acute Type B will include:

- a) Paraplegia
- b) Visceral malperfusion
- c) Renal malperfusion
- d) Ischaemic limbs
- e) Contained leak
- f) Enlarging pleural effusions

Other issues:

- a) Retrograde dissection evolving into an acute Type A
- b) Persistent pain
- c) Rapid expansion
- d) Uncontrolled hypertension

Under these circumstances intervention may include TEVAR, Arch and Frozen Elephant Trunk or as a last resort open thoracoabdominal aortic surgery.

## **Referral**

Once the patient has been accepted into RLUH or LHCH through pathways described above, coordination is required with the Matron in-charge of the relevant Critical Care Area. Transfers should not occur until a bed is confirmed.

## **Transfer**

Transfers should be with a medical escort and include:

- a) IV access and strict BP control with Labetalol
- b) Arterial line
- c) Urinary catheter

## **Virtual MDT Management**

Management of these patients will typically involve a virtual MDT of relevant specialists to agree and ensure appropriate management.

## **Aortic MDT**

All cases are mandated to undergo a formal MDT discussion at monthly meetings at LHCH on second Wednesday of every month at 0815 in Radiology Seminar Room. Any interested Physician is welcome to attend.

## **Outpatient Reviews**

Weekly clinics in LHCH or RLUH are available to review patients in follow-up phase. Follow-up will likely be for life and relevant referrals are made including genetic service.

## **Standard Setting**

### **Key Quality Markers**

**1) Door to treatment decision (CCA or Theatre) within 6 hours**

**2) Door to Diagnosis within 4 hours of arrival**

**3) Diagnosis to treatment decision within 2 hours**

**4) Early management complicated and uncomplicated dissections**

a. All the following:

i. Critical care bed

ii. Labetalol infusion

iii. Arterial line

iv. Catheter

v. IV access

**5) Audit trail and review of time lines from treatment decision (medical versus intervention) to destination**

### **Feedback**

Data on the pathway will be fed back on an annual basis.



### **3) Acute on chronic thoracoabdominal disease**

Acute on chronic thoracoabdominal disease is a common referral and represents the far end of complexity in terms of management. Depending on the age, co-morbidities and general fitness the patient may be transferred into LHCH or RLUH. Diagnosis, early management and referral should follow the same guidelines above for acute Type B aortic syndromes. Aortic MDT discussion is central to bespoke patient specific management.

#### **4) Other (Mycotic aneurysms, Vasculitis, Trauma)**

##### **1) Mycotic aneurysm**

- a. Mycotic or infected aneurysms are complex pathologies and difficult to manage. Referrals should be via LHCH or RLUH depending on proximity and pathology. Transfer is often warranted.

##### **2) Active vasculitis**

- a. Active vasculitis of large vessels is complex to manage and often multi-disciplinary with our physicians. Referrals should be via LHCH or RLUH depending on proximity and pathology. Transfer is often warranted.

##### **3) Blunt Traumatic Aortic Rupture**

- a. BTAR requires immediate referral to the on-call consultant at LHCH or RLUH.

## **Novel treatments, best medical management and exiting the pathway**

Although aortic emergencies often affect young people, a significant percentage of presentations are in very elderly patients with multiple co-morbidities. For such patients, particularly acute on chronic thoracoabdominal aortic disease, the most appropriate strategy may be “Best Medical Care” in a critical care environment or palliation either in hospital, local hospice or home. A few select few elderly patients presenting with acute Type A aortic dissection may be eligible for novel endovascular intervention.

### **Additional Information**

- 1) American Heart Association Guidelines
- 2) European Guidelines

