Acute Aortic Syndrome Pathway: *recommendations for diagnosis, early management, referral and transfer within North West of England, North Wales and Isle of Man.*

Short title: NW Acute Aortic Pathway

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6) NW Response to Coroners Regulation 28
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1) Background to national service reconfiguration

Acute aortic dissection is a life-threatening emergency requiring highly specialised care. Early diagnosis and treatment are key to a successful outcome. Chest pain is the principal symptom and therefore diagnosis is often delayed due to the rarity of this disease compared to the more common diseases presenting with chest pain such as acute coronary syndromes and pulmonary embolism (Appendix: Delayed Recognition of Acute Aortic Dissection, HSIB, 2017). A key publication in 2017 highlighted unwarranted variation in the quality of care for patients with diseases of the thoracic aorta in the UK.

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ORIGINAL RESEARCH

Unwarranted Variation in the Quality of Care for Patients With Diseases of the Thoracic Aorta

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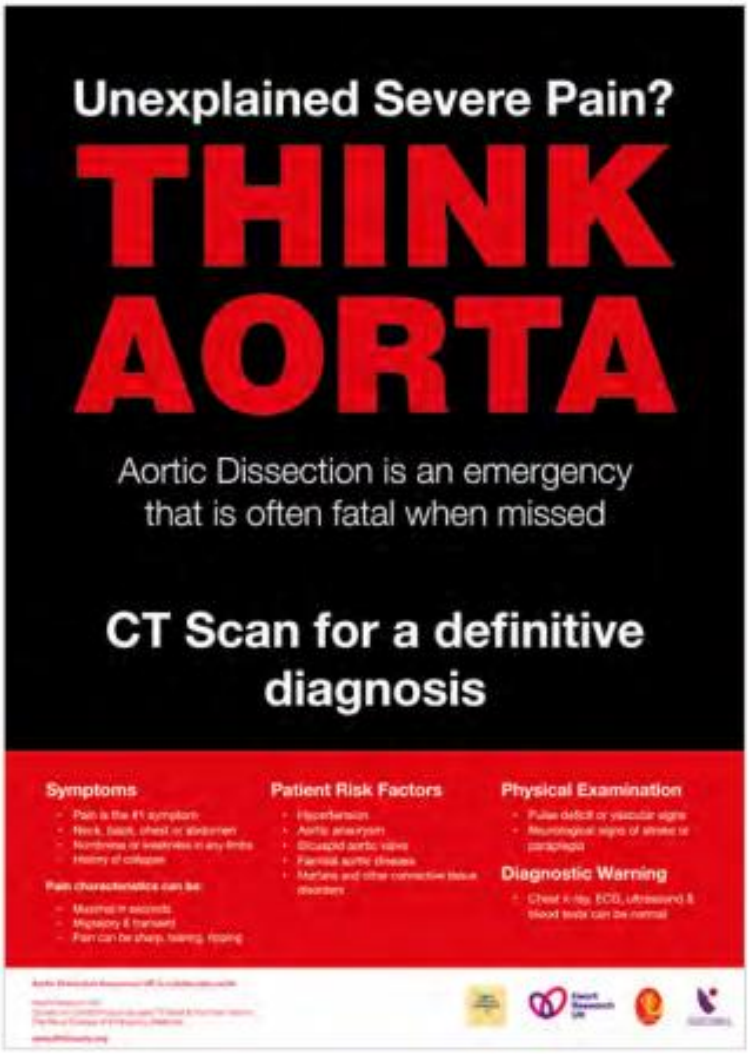
Several campaigns have been launched to raise awareness in acute medical facilities including "*Thinks Aorta*":



Adding to the complexity is the shared responsibility for managing these patients including:

1) Vascular Surgery
2) Cardiac Surgery
3) Interventional Radiology
4) Cardiology
5) General Medicine
6) Intensive Care

In 2022, the NHS published a document describing the "Aortic Dissection Toolkit", a nationwide initiative to set up regional networks to improve diagnosis, transfer, and treatments for these conditions (Appendices: Acute Aortic Dissection Pathway Toolkit V1.0, AAD Toolkit Interactive, AAD Self-Assessment Questionnaire, SCIP document). The toolkit has 7 Principles.

Principle 1 requires a Standard Operating Procedure for each of the regional network and is the focus of this document.

The North-West Region:

Within the North-West of England, which also takes referrals from the North of Wales and The Isle of Man, are 4 cardiac surgery centres:

1) Blackpool Victoria Hospital NHS Trust
2) Manchester Royal Infirmary NHS Trust
3) Manchester Wythenshawe Hospital NHS Trust
4) Liverpool Heart and Chest Hospital NHS Trust

Vascular Surgery and Interventional Radiology as they relate to aortic disease are:

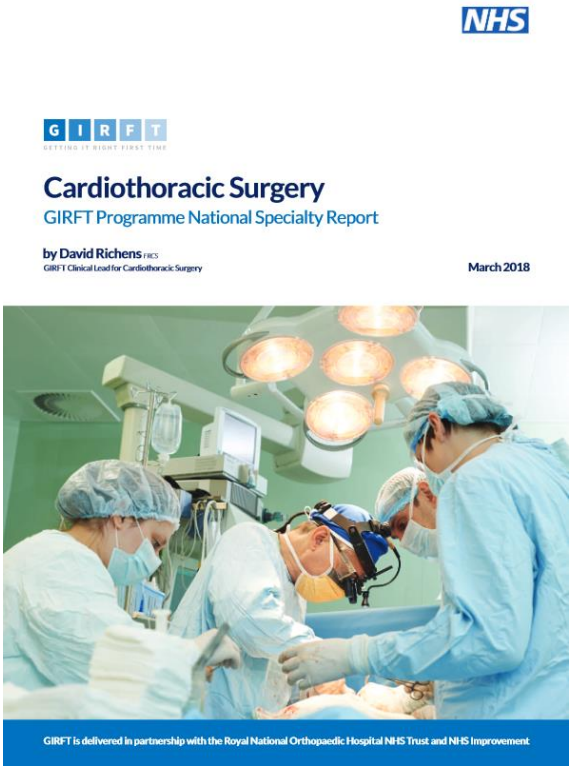
1) Liverpool University Hospital Foundation Trust – Aintree University Hospital.
2) Manchester University Hospital Foundation Trust – Manchester Royal Infirmary NHS Trust.
3) Lancashire Teaching Hospitals NHS Foundation Trust – Preston.

Existing policies:

A Coroners Regulation 28 was issued against Liverpool, Manchester and Blackpool/Preston hospitals following the death of a patient in 2019, which resulted in an agreement between the Trusts on how to manage acute aortic emergencies (Appendix). This document and pathway are currently operational but does not address all aspects of the Aortic Dissection Toolkit. A separate policy document has been published by Blackpool/Preston Hospitals (Appendix) and MRI/Wythenshawe Hospitals (Appendix) which largely follows the regional pathway.

Levers to change:

The Cardiac GIRFT (Appendix) is very prescriptive around the changes which are felt necessary in the management of patients with thoracic aortic disease.



GIRFT "Aortic" Recommendations

- GIRFT has recommended (Recommendation 14): "*Ensure that aortic syndrome patients are only operated upon by rotas of acute aortic syndrome specialist teams*". This is based on their analysis of the UK national data set (HES and NICOR) showing high volume aortovascular surgery is associated with better outcomes.
- Recommendation 14 is broken down into:
- **14a** Create rotas of specialist surgeons allied to networks of referring hospitals to cover geographic areas.
- **14b** Ensure that all surgeons on the rota meet minimum activity thresholds as defined by 14c.
- **14c** Define minimum activity thresholds for the surgeons.
- **14d** Establish formal agreements between referring hospitals, receiving specialists' units and ambulance service for transfer of AAD patients to the relevant specialist centre. The

arrangements should include a dedicated phone number for referrals and service coordination.

The Vascular GIRFT has little comment on acute aortic disease.

This document presents basic information for the non-specialist on diagnosis, management, and transfer of patients with acute aortic syndromes in the North-West of England, North Wales, and Isle of Man.

2) Acute Aortic Syndromes (AAS) explained: a spectrum of disease

Acute Aortic Syndromes represent a spectrum of disease analogous to the acute coronary syndromes.

a. *Type A Aortic Dissection*

b. *Type B Aortic Dissection*

    a. Uncomplicated dissection

    b. Complicated dissection

c. *Acute on chronic thoracoabdominal aortic disease*

a. Type A aortic dissection

Pathology

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

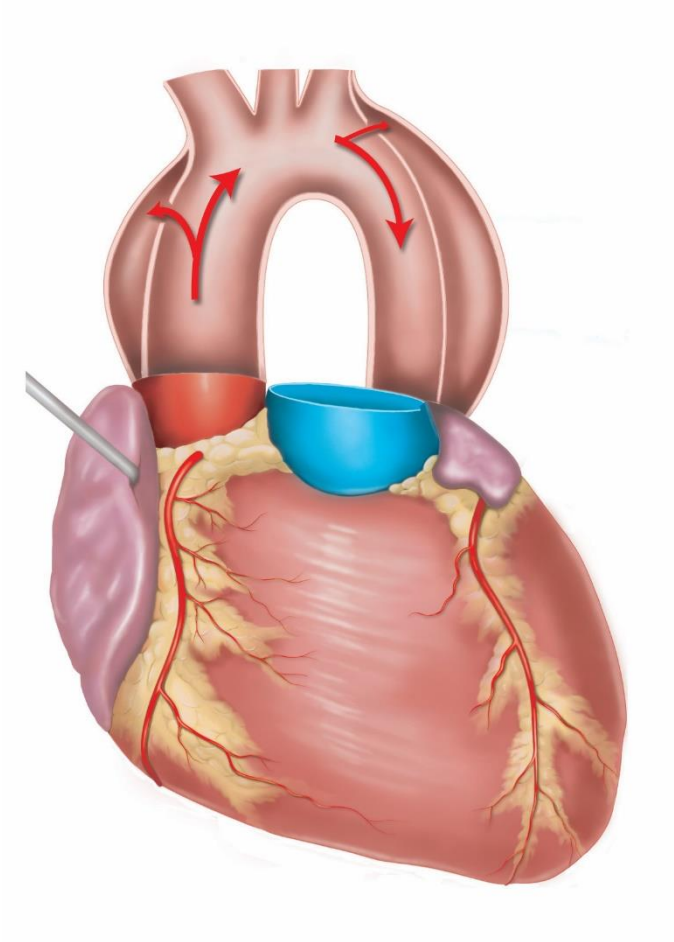


Figure: *Acute Type A involves the ascending aorta while acute Type B does not.*

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.

b. Type B aortic 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 – so-called complicated disease. Management of the Type B AAS's is further complicated by the equivocal evidence base around TEVAR in uncomplicated but high risk disease.

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 specialised centres.

c. Acute on chronic thoracoabdominal aortic disease

Pathology

Chronic aneurysms of the entire aorta are vulnerable to acute events such as dissection, IMH and rupture.

3) NW Standard Operating Policy for acute aortic dissection

(Printable stand-alone sub-section for clinical use)

North-West Region clinical advice for managing patients with Acute Aortic Syndromes

(Standard Operating Policy)

Policy document: *North-West Aortic Dissection Toolkit 2023*

a. Emergency contact details:

Liverpool

(Liverpool Heart and Chest Hospital (LHCH) and Aintree University Hospitals (LUHFT – Liverpool University Hospital Foundation Trust))

Type A and B referrals (LHCH)

- LHCH Switch Tel: 0151 2281616
- On-call middle grade contact:
  - Aortic Fellow (Mobile phone via Switch)
  - Cardiac SpR on-call (Bleep 2767 via Switch)
- On-call Consultant Aortic Surgeon (Mobile phone via Switch)
- Clinical Lead Aortic Surgery (Mark Field)

Trauma referrals (Aintree University Hospital (LUHFT)

- LUHFT Switch Tel: 01515255980
- On-call middle grade contact:
  - Vascular SpR on-call (Bleep via Switch)
- On-call Consultant Vascular Surgeon (Mobile phone via Switch)
- Clinical Lead Vascular Surgery (Rob Fisher)

Blackpool

(Blackpool Victoria Hospital (BVI) and Preston Hospital)

Type A and Type B referrals (Blackpool Victoria Hospital)

- Blackpool Switch Tel: 0151 2281616
- On-call middle grade contact:
  - Cardiac SpR on-call (Bleep via Switch)
- On-call Consultant Aortic Surgeon (Mobile phone via Switch)
- Clinical Lead Aortic Surgery (Nidal Bittar)

Trauma referrals (Preston Hospital)

- Preston Switch Tel 01772716565
- On-call middle grade contact:
  - Vascular SpR on-call (Bleep via Switch)
- On-call Consultant Vascular Surgeon (Mobile phone via Switch)
- Clinical Lead Vascular Surgery

Manchester

(Manchester Royal Infirmary/Manchester Wythenshawe Hospital, MUHFT)

Type A (MRI and Wythenshawe)

- MRI/Wythenshawe (MUHFT) Switch Tel: 0151 2281616
- On-call middle grade contact:
    - Cardiac SpR on-call (Bleep via Switch)
- On-call Consultant Aortic Surgeon (Mobile phone via Switch)
- Clinical Lead Aortic Surgery (Paul Waterworth)

Type B (MRI and Wythenshawe)

- MRI/Wythenshawe (MUHFT) Switch Tel: 0151 2281616
- On-call middle grade contact:
    - Vascular SpR on-call (Bleep via Switch)
- On-call Consultant Vascular Surgeon (Mobile phone via Switch)
- Clinical Lead Vascular Surgery (Jonathan Gosh)

Trauma referrals (MRI)

i. MRI Switch Tel:
ii. On-call middle grade contact
    i. Vascular SpR on-call (Bleep via Switch)
iii. On-call Consultant Vascular Surgeon (Mobile phone via Switch)
iv. Clinical Lead Vascular Surgery (Jonathan Gosh)

North-west England, North Wales and Isle of Man
Management of *Acute Aortic Dissection*: *Quick reference wall chart*

THE AORTIC DISSECTION CHARITABLE TRUST

Unexplained chest pain?
"Think Aorta"
⇩
CT Whole Aorta

Type A aortic dissection

Medical management:

- IV access
- Labetalol infusion
- Urinary catheter
- Arterial line
- Oxygen
- Analgesia

Target BP: Below 120mmHg

Identify high risk features to referrer:

- Uncontrolled BP (>120mmHg)
- Peri-arrest
- Obvious cardiac tamponade
- Evidence of myocardial infarction
- Evidence of general low cardiac output and/or malperfusion (lactate >4)
- Evidence of limb ischaemia, gut ischaemia or stroke

Safe versus timely transfer:

- Medical escort with controlled BP.
- Do not "scoop and run"

Discuss transfer arrangement with receiving centre

Type B aortic dissection

Medical management in Critical Care environment

- IV access
- Labetalol infusion
- Urinary catheter
- Arterial line
- Oxygen
- Analgesia

Target BP: Below 120mmHg

Identify "Complicated" acute Type B dissection:

- Paraplegia
- Visceral and renal malperfusion
- Ischaemic limbs
- Enlarging pleural effusions
- Persistent pain
- Rapid expansion
- Retrograde Type A
- Uncontrolled BP

- Patients with Complicated acute Type B aortic dissection will require safe and timely transfer
- Patients with Uncomplicated acute Type B aortic dissection will be managed in referring Centre with remote support

Liverpool	Manchester	Blackpool
• 0151 2281616 (LHCH)	• 0300 3309444 (MRI/Wythenshawe)	• 01253 300000 (BVH)
• Aortic Fellow (Switch) • Cardiac Surgery Registrar (Switch)	• Cardiac Surgery Registrar for Type A (Switch) • Vascular Surgery Registrar for Type B (Switch)	• Cardiac Surgery Registrar (Switch)
• On-call Aortic Surgeon (Switch) • Clinical Leads: Prof Field and Prof Torella	• On-call Aortic and Vascular Surgeon (Switch) • Clinical Leads : Mr Waterworth & Mr Murry	• On-call Aortic Surgeon (Switch) • Clinical Leads: Mr Bittar & Miss Drinkwater

NHS Liverpool Heart and Chest Hospital NHS Foundation Trust
NHS Lancashire Teaching Hospitals NHS Foundation Trust
NHS Manchester University NHS Foundation Trust
NHS Liverpool University Hospitals NHS Foundation Trust

Full details may be found at:

Type A aortic dissection management and referral process

i. Presentation, Management, and Referral process

Our approach is based around the American Heart Association Guidelines 2022 (Appendix).

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 pain, 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.

Recommendations for AAS: Diagnostic Evaluation (Imaging, Laboratory Testing)

COR	LOE	RECOMMENDATIONS
1	C-LD	1. In patients with a suspected AAS, CT is recommended for initial diagnostic imaging, given its wide availability, accuracy, and speed, as well as the extent of anatomic detail it provides.[1-5]
2a	C-LD	2. In patients with a suspected AAS, TEE and MRI are reasonable alternatives for initial diagnostic imaging.[1-6]

Management

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

1) IV access

2) Labetalol infusion

3) Urinary catheter

4) Arterial line

5) Oxygen

6) Analgesia

Target BP: Below 120mmHg

This is important to reduce risks of rupture or extension of the dissection as well as allow safe transfer. This broadly follows guidance from American Heart Association.

Recommendations for Acute Medical Management of AAS
Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients presenting to the hospital with AAS, prompt treatment with anti-impulse therapy with invasive monitoring of BP with an arterial line in an ICU setting is recommended as initial treatment to decrease aortic wall stress.[1-5]
1	C-LD	2. Patients with AAS should be treated to an SBP <120 mm Hg or to lowest BP that maintains adequate end-organ perfusion, as well as to a target heart rate of 60 to 80 bpm.[3,6]
1	B-NR	3. In patients with AAS, initial management should include intravenous beta blockers, except in patients with contraindications.[2,5,7]
2a	B-NR	In those with contraindications or intolerance to beta blockers, initial management with an intravenous non-dihydropyridine calcium channel blocker is reasonable for heart rate control.[1,2,5]
1	C-LD	4. In patients with AAS, initial management should include intravenous vasodilators if the BP is not well controlled after initiation of intravenous beta-blocker therapy.[8]
1	C-EO	5. Patients with AAS should be treated with pain control, as needed, to help with hemodynamic management.

Onward management for acute Type A aortic dissection/IMH is principally surgery. Within the North-West, North Wales and IOM, this would be within one of the 4 centres for cardiac surgery (Blackpool Royal Infirmary, Manchester Royal Infirmary, Wythenshawe Hospital or Liverpool Heart and Chest Hospital).

Recommendations for Initial Surgical Considerations in Acute Type A Aortic Dissection
Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients presenting with suspected or confirmed acute type A aortic dissection, emergency surgical consultation and evaluation and immediate surgical intervention is recommended because of the high risk of associated life-threatening complications.[1,2]
2a	B-NR	2. In patients presenting with acute type A aortic dissection, who are stable enough for transfer, transfer from a low- to a high-volume aortic center is reasonable to improve survival.[3,4]
2a	B-NR	3. In patients presenting with nonhemorrhagic stroke complicating acute type A aortic dissection, surgical intervention is reasonable over medical therapy to reduce mortality and improve neurologic outcomes.[5,6]

The North-West have agreed to maintain 3 separate rotas of specialist aortic surgeons in Blackpool, Liverpool, and Manchester. Acute patients should be referred to their local and or traditional centres of cardiac surgery along which elective patients flow. Additional aspects of the emergency referral process for acute Type A disease:

1) All referrers should refer to their local centre for cardiac surgery.
2) Receiving centres are in the main not allowed to turn down patients for life saving surgery due to bed capacity alone (Critical

Care Network Memorandum of Understanding, Appendix). Patients may be taken directly to theatre, bypassing Critical Care.

3) Receiving centres unable to accept a patient, due to unavailability of a theatre team (ie transplant or other emergency in progress) or perceived complexity of the case (ie concurrent malperfusion, concurrent arch and thoracoabdominal disease), should take charge of finding a suitable destination. This is not the responsibility of the acute medical care facility in which the patient is admitted. In principle, any such transfers should be agreed at consultant-to-consultant level.

4) Whilst the AHA guidelines suggest transfer of patients diagnosed with acute Type A aortic dissection to high volume centres if stable, all referring centres in the North-West have roughly equal volumes and outcomes.

2. In patients presenting with acute type A aortic dissection, who are stable enough for transfer, transfer from a low- to a high-volume aortic center is reasonable to improve survival.[3,4]

Type A Referral and transfer process by Centre (Cardiac Surgery)

Liverpool Heart and Chest Hospital

i) Referral

All patients with acute Type A aortic pathologies in North Wales, Merseyside, West Cheshire, and Isle of Mann should be referred to LHCH. At the Liverpool Heart and Chest Hospital there are a number of ways to refer a patient 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, Professor Field, Miss Harrington, Mr Nawaytou, Mr Othman or Mr Kenawy.

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.

ii) Transfer

Once accepted by the surgeon, there will need to be a discussion between surgeon and our on-call Intensivist regarding an ITU bed. Availability of an ITU bed however should not prevent a transfer as the patient may be brought directly to theatre. 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 arrangement.

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. Equally, transfers need to be timely and if delays are anticipated in a transfer, then a discussion with the on-call surgeon should take place.

iii) 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 where no ITU bed is available for admission and assessment or those identified at very high risk:

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

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.

Blackpool Royal Infirmary

i) Referral

All patients with acute Type A aortic pathologies in South- West Lancashire should be referred to BVH. At the Blackpool Victoria Hospital there are a number of ways to refer a patient however the fastest pathway is to ring switchboard (xxxx) and ask to be put through to the Cardiac Surgery Registrar. The Registrar will take the details and request transfer of imaging CT scan. 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…..

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.

iv) Transfer

Once accepted by the surgeon, there will need to be a discussion between surgeon and our on-call Intensivist regarding an ITU bed. Referrer to arrange a time-critical ambulance transfer by calling '999' and requesting a 'blue light and sirens' ambulance with a paramedic crew to immediately transfer patient to receiving hospital. Availability of an ITU bed however should not prevent a transfer as the patient may be brought directly to theatre. 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 arrangement.

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. Equally, transfers need to be timely and if delays are anticipated in a transfer, then a discussion with the on-call surgeon should take place.

iii)Destination within BVH

Patient will normally be transferred into the Critical Care Area which may be:1) Intensive Care, …….For some selected patients the destination will be directly through the Critical Care Area into the Theatre Suit. These patients will be those where no ITU bed is available for admission and assessment or those identified at very high risk:

- Peri-arrest
- Obvious tamponade
- Evidence of myocardial infarction

- Evidence of general low cardiac output and/or malperfusion (lactate >4)
- 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.

i) Referral

All patients with acute Type A aortic pathologies in Greater Manchester Area should be referred to the Cardiac Surgeon on-call for Aortic Disease via MUHFT Switchboard, asking for on-call SpR for Cardiothoracic Surgery. The SpR will take the details and request transfer of imaging CT scan. If the SpR cannot be contacted, then the consultant surgeon on-call should be contacted directly via switchboard. The Aortic on-call team currently includes: Mr Waterworth, Mr Mehta….. The destination for surgery may be MRI or Wythenshawe Hospital depending on the rota.

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.

a. Transfer

Once accepted by the surgeon, there will need to be a discussion between surgeon and our on-call Intensivist regarding an ITU bed at WTWA or MRI. Referrer to arrange a time-critical ambulance transfer by calling '999' and requesting a 'blue light and sirens' ambulance with a paramedic crew to immediately transfer patient to receiving hospital.

Availability of an ITU bed however should not prevent a transfer a the patient may be brought directly to theatre. 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 arrangement.

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. Equally, transfers need to be timely and if delays are anticipated in a transfer, then a discussion with the on-call surgeon should take place.

b. Destination within Manchester Hospitals

Patient will normally be transferred into:

- Cardiac Intensive Care Unit (CICU at MRI or CTCCU at Wythenshawe) for Type A Dissection

For some selected patients the destination will be directly through the Critical Care Area into the Theatre Suit. These patients will be those where no ITU bed is available for admission and assessment or those identified at very high risk:

- Peri-arrest
- Obvious tamponade
- Evidence of myocardial infarction
- Evidence of general low cardiac output and/or malperfusion (lactate
- >4)
- 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.

ii)End of Life Care

End of life care should happen in the patient's local hospital, close to family and friends, with support from the local palliative care team. A palliative approach to care will provide a dignified death. This includes pain relief, anxiety relief, maximising comfort, and support from family and/or carers.

iii)Standard Setting (KQI)

The NW hospitals have agreed a set of Key Quality Indicators with which to benchmark process and allow audit and monitor improvement. These will be slightly different when the patient is in a remote facility requiring transfer compared to an in-house facility.

Key Quality Indicators:

i) Out with referring hospital:

1) Door to skin within 8 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 within 2 hours

5) Theatre

a. Arrival to knife to skin within 2 hours

ii) Within admitting hospital

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) Theatre

a. Arrival to knife to skin within 2 hours

iv)Feedback to referring centres in the network:

Feedback to referring A&E/AMU's will be on an annual basis to the Head of Department and benchmarked against our KQI. Data acquisition will require bespoke prospective/retrospective audits.

v)MDT arrangements:

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

vi)Follow-up:

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

Type B aortic dissection management and referral process

a. Uncomplicated dissection

b. Complicated dissection

i) Presentation, Management and Referral Process

Patients present is 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.

Recommendations for AAS: Diagnostic Evaluation (Imaging, Laboratory Testing)

COR	LOE	RECOMMENDATIONS
1	C-LD	1. In patients with a suspected AAS, CT is recommended for initial diagnostic imaging, given its wide availability, accuracy, and speed, as well as the extent of anatomic detail it provides.[1-5]
2a	C-LD	2. In patients with a suspected AAS, TEE and MRI are reasonable alternatives for initial diagnostic imaging.[1-6]

ii) 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 mostly be managed medically and with serial imaging typically in their presenting hospital with the advice of the specialist team. 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
4) Arterial line
5) Oxygen
6) Analgesia

Target BP: Below 120mmHg, however in some patients with malperfusion, "permissive hypertension" may be acceptable.

This is important to reduce risks of rupture or extension of the dissection as we ll as allow safe transfer. This broadly follows guidance from American Heart Association.

Recommendations for Acute Medical Management of AAS
Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients presenting to the hospital with AAS, prompt treatment with anti-impulse therapy with invasive monitoring of BP with an arterial line in an ICU setting is recommended as initial treatment to decrease aortic wall stress.[1-5]
1	C-LD	2. Patients with AAS should be treated to an SBP <120 mm Hg or to lowest BP that maintains adequate end-organ perfusion, as well as to a target heart rate of 60 to 80 bpm.[3,6]
1	B-NR	3. In patients with AAS, initial management should include intravenous beta blockers, except in patients with contraindications.[2,5,7]
2a	B-NR	In those with contraindications or intolerance to beta blockers, initial management with an intravenous non-dihydropyridine calcium channel blocker is reasonable for heart rate control.[1,2,5]
1	C-LD	4. In patients with AAS, initial management should include intravenous vasodilators if the BP is not well controlled after initiation of intravenous beta-blocker therapy.[8]
1	C-EO	5. Patients with AAS should be treated with pain control, as needed, to help with hemodynamic management.

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 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 and considered for TEVAR in the sub-acute phase (2-12 weeks).

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 a specialist centre (Preston, MRI or LHCH).

Complicated acute Type B will include:

a) Paraplegia
b) Visceral malperfusion
c) Renal malperfusion (with resistant hypertension)
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. Complex open surgery under these circumstances should be referred to LHCH.

Type B referral and transfer process by Centre

Liverpool Heart and Chest Hospital

i) Referral

All patients with complicated acute Type B aortic pathologies in North Wales, Merseyside, West Cheshire, and Isle of Mann should be referred to LHCH. At the Liverpool Heart and Chest Hospital there are a number of ways to refer a patient 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, Professor Field, Miss Harrington, Mr Nawaytou, Mr Othman or Mr Kenawy. The team will involve Vascular Surgery and Interventional Radiology as required.

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.

ii) Transfer

Once accepted by the surgeon, there will need to be a discussion between surgeon and our on-call Intensivist regarding an ITU bed.

Availability of an ITU bed however should not prevent a transfer as the patient may be brought directly to theatre. 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 arrangement.

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. Equally, transfers need to be timely and if delays are anticipated in a transfer, then a discussion with the on-call surgeon should take place.

iii) 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.

BVH and Preston Hospital

i) Referral

All patients with complicated acute Type B aortic pathologies in South Lancashire Area should be referred to the Cardiac Surgeon on-call for Aortic Disease via BVH Switchboard, asking for on-call Cardiothoracic SpR.

This pathway has been developed for the following Trusts:

• Lancashire Teaching Hospital NHS Foundation Trust (LTHTR).

• Wrightington, Wigan and Leigh NHS Foundation Trust (WWL).

• Blackpool Teaching Hospital NHS Foundation Trust (BVH).

• Royal Lancaster Infirmary (RLI).

• University Hospital of Morecombe Bay NHS Foundation Trust (UHMB).

• Westmorland General Hospital (WGH

The SpR will take the details and request transfer of imaging CT scan. If the SpR cannot be contacted, then the consultant surgeon on-call should be contacted directly via switchboard.

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.

ii) Transfer

Once accepted by the cardiac surgeon, there will need to be a discussion between cardiac surgeon and vascular surgeon/interventional radiologist as well as on-call Intensivist regarding an ITU bed at Preston or Blackpool.

Availability of an ITU bed however should not prevent a transfer as the patient may be brought directly to theatre. 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 arrangement.

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. Equally, transfers need to be timely and if delays are anticipated in a transfer, then a discussion with the on-call surgeon should take place.

iii) Destination within Preston and BVH

Patient will normally be transferred into a critical care bed at either hospital dependent on need and availability.

i) Referral

All patients with complicated acute Type B aortic pathologies in Greater Manchester Area should be referred to the Vascular Surgeon on-call for Aortic Disease via MUHFT Switchboard, asking for on-call SpR for Vascular Surgery. The SpR will take the details and request transfer of imaging CT scan. If the SpR cannot be contacted, then the consultant surgeon on-call should be contacted directly via switchboard. On occasions, referral may be to the on-call cardiac surgeon.

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.

ii) Transfer

Once accepted by the surgeon, there will need to be a discussion between surgeon and our on-call Intensivist regarding an ITU bed at WTWA or MRI.

Availability of an ITU bed however should not prevent a transfer as the patient may be brought directly to theatre. 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 arrangement.

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. Equally, transfers need to be timely and if delays are anticipated in a transfer, then a discussion with the on-call surgeon should take place.

a. Destination within Manchester Hospitals

Patient will normally be transferred into:

- Cardiac Intensive Care Unit (CICU at MRI or CTCCU at Wythenshawe) for Type A Dissection

ii)End of Life Care

End of life care should happen in the patient’s local hospital, close to family and friends, with support from the local palliative care team. A palliative approach to care will provide a dignified death. This includes pain relief, anxiety relief, maximising comfort, and support from family and/or carers.

iii)Standard Setting (KQI)

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

IV.Feedback

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

v.Virtual MDT Management

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

vi.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.

vi.Follow-up

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.

4)Additional nuances

a)Intramural Haematoma:

For the purposes of referring medical teams, IMH should be managed as per a frank acute aortic dissection (Type A and B).

b)Penetrating Aortic Ulcer (PAU):

PAU is typically found in the thoracobabdominal aorta and aortic arch. Rarely in the ascending aorta. Patients should be managed as per protocols for acute Type A or Type B aortic dissections.

c)Acute on chronic thoracoabdominal aortic disease:

Acute aortic syndromes on chronic thoracoabdominal disease is a complicated presentation requiring highly specialised teams. Patients should be referred along the Type B pathway and medical teams will arrange onward referral as required.

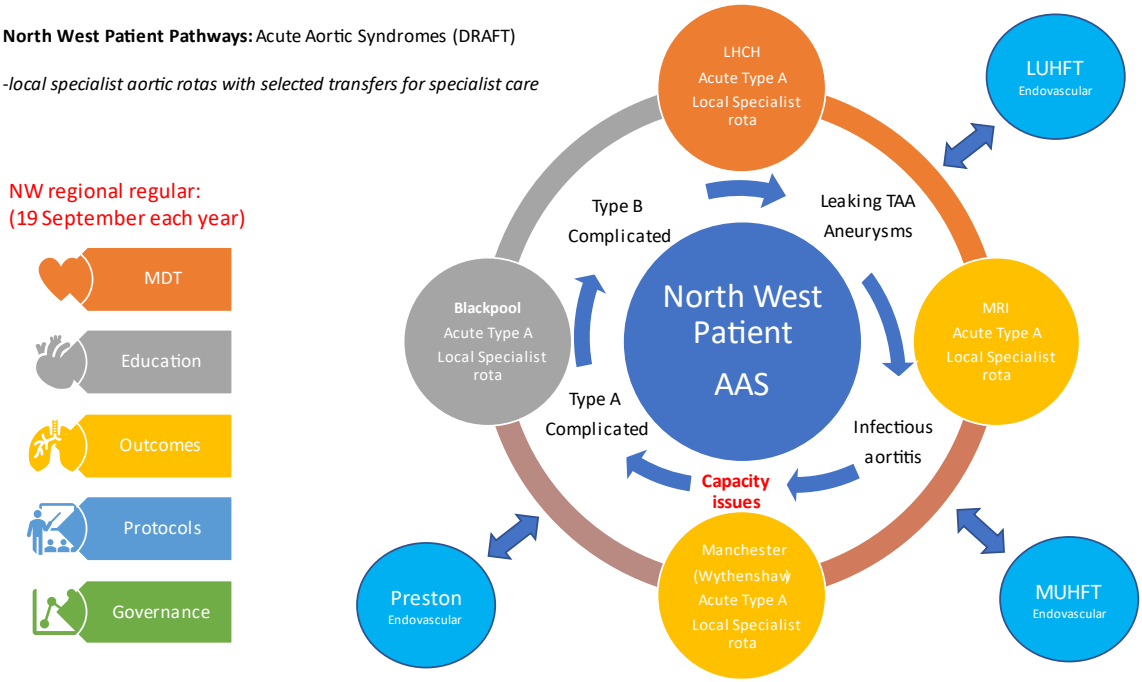
d)Transfers, Complexity and capacity issues:

Transfer of patients between centres in the North-West, outside the guidance above, may be indicated as follows, and only with consultant-to-consultant discussion:

1)Wythenshawe hospital are involved in a transplant and have no available theatre team

2)Teams are already engaged in emergency surgery

It is not appropriate to transfer patients on the basis of "no aortic surgeon available" without consultant-to-consultant discussion. In addition, the unavailability of an ITU bed is not an appropriate reason to decline a patient immediately life saving intervention. When patients lay outside of centres, it is not the responsibility of that A&E or MAU to find alternate centres but rather the centre to which the primary phone call is made.



5)Trauma

Blunt traumatic aortic injury requires immediate input from Vascular Surgery with a view to TEVAR, working with a Poly-Trauma Team.

Liverpool – Refer to Vascular Team at University Hospital Aintree

Manchester – Refer to Vascular Team at MRI

Blackpool/Prestion – Refer to Vascular Team at Preston