Proposed Aortic Surveillance Flowchart

Patient has a dilated thoracic aorta identified on imaging (e.g. Echo, thoracic CT)[1]

Clinical assessment – age, family history, comorbidities and frailty, aortic valve morphology and function, BP.

Ensure modifiable risk factors (hypertension) are treated

Aortic dilatation is common. Further assessment is recommended if patient would be considered for prophylactic aortic surgery plus any of the following[2]:

- FHx of aortic dilatation/dissection
- Bicuspid aortic valve
- Age <60y
- Absolute aortic dimension >40mm

Is the patient felt likely to benefit from surveillance?[3]

High suspicion of familial aortopathy / criteria met for genetic testing

Referral to Genetics / Family Screening Service

YES

NO

Refer for MRI or CT Aorta[5]

MEETS SURGICAL CRITERIA

ABNORMAL

NORMAL

Dilated segment well-seen on echo, no other aortopathy

YES

Echo surveillance – further CT/MRI only at clinician request

NO

No further action required

MRI or CT Aorta at 1 year to assess for rapid progression

MEETS SURGICAL CRITERIA

RISK STRATIFICATION (see table)

Low Risk 5 Years [6]

Moderate Risk 3 Years [6]

High Risk Annual [6]

CONSIDER EHCP / "DISSECTION PLAN"[7]

Patient has become no longer suitable for surveillance (e.g. advancing frailty)[8]

Surveillance MRI / CT scans at intervals dictated by risk group

Patient becomes pregnant / plans pregnancy[9]

-Referral to Cardio-obstetric clinic
-Updated baseline MRI Aorta and repeated during pregnancy / puerperium (under expert guidance)
-Some patients should be counselled against pregnancy
-Surgical criteria may differ
-Return to standard risk-based surveillance post-puerperium

MEETS SURGICAL CRITERIA

Clinical assessment & referral for intervention[10]

POST-OP SURVEILLANCE >1y

Aortic Surveillance Risk Groups

This table is not exhaustive and certain factors (e.g. particularly high-risk family history or rapid rate of progression in aortic size) may prompt change in risk category or referral for intervention.

Low Risk	Moderate Risk	High Risk	Surgical Criteria
Trileaflet valve with aorta 40-44mm	Trileaflet valve with aorta 45-49mm	Aorta 50-54mm – any aetiology not meeting surgical criteria	Aorta ≥ 55mm – any aetiology
	Bicuspid valve with aorta ≤ 44mm	Bicuspid valve with aorta 45-49mm, or with >3mm/yr increase	Aorta ≥ 50mm with Bicuspid aortic valve & risk factors*, **OR** Marfan's syndrome **OR** Type 4 EDS
		Familial Aortopathies (Marfans, Loeys-Dietz, Type 4 EDS) of any aortic dimensions not meeting surgical criteria	Aorta ≥ 45mm with Loeys-Dietz Syndrome **OR** Marfan's & risk factors† **OR** Bicuspid valve & need for AVR.
Turner's Syndrome with normal aortic dimensions (size index <20mm/m^2) and no risk factors‡	Turner's Syndrome with normal aortic dimensions (size index <20mm/m^2) & any risk factor‡	Turner's Syndrome with size index 20-24mm/m^2	Turner's Syndrome with aortic size index >25mm/m^2 & risk factors‡
	Post-op thoracic aneurysm repair, without Familial Aortopathy, with stable post-op dimensions at 1/6/12 months	Post-op thoracic aneurysm repair with Familial Aortopathy. All post-op Aortic Dissection	Post-op residual type B dissection with descending aorta ≥ 60mm or malperfusion **OR** significant post-op complication e.g. pseudoaneurysm

***Bicuspid Valve** & Coarctation of the aorta, systemic hypertension, family history of dissection, or increase in aortic diameter >3 mm/year

† Marfan's Syndrome & Family history of AD and/or aortic size increase >3 mm/year, Severe AR or MR, Desire for pregnancy

‡ Turner's Syndrome & bicuspid valve, elongation of the transverse aorta, Coarctation, or hypertension

Footnotes

1 There is considerable similarity of the normal ranges between the published normal ranges for MRI (Burman et al, Davis et al), Echo (NORRE dataset, used in BSE guidelines) and CT (Lim et al and the Copenhagen dataset), particularly for the upper limit of normal at the Sinus of Valsalva, and it is reasonable to assume that a dilated aorta by any modality is potentially suitable for further assessment by cross-sectional imaging.

2 It is recognised that there are currently no data addressing outcomes in patients with very mild dilatation only, and certainly no randomised trials evaluating any specific criteria or surveillance strategy. Given the most recent British Society of Echo guidelines advise aortic measurements normalised to the patient's height, it is now more common to find patients who do have aortic dimensions outwith the normal population values in large datasets (e.g. NORRE), but still have relatively small (<40mm) absolute aortic size. Local audit data at NUTH found that 28% of all echocardiograms (i.e. performed in a population with a high prevalence of disease, rather than the general population) had abnormal aortic root dimensions by BSE/NORRE criteria.

Further aortic imaging of more than a quarter of all patients undergoing echocardiography would not be feasible, hence the desire to target further assessment on those most likely to benefit. These specific criteria for proceeding with further aortic assessment are consensus-based, and were developed between clinicians at North Tees & Hartlepool, South Tees Hospitals and Newcastle Upon Tyne Hospitals after presentation of the available evidence and local audit data.

3 Entry into surveillance is a clinical decision based on likelihood of the patient going on to derive benefit, were their aortopathy to progress. Patients who would not be surgical candidates (or would never wish for prophylactic surgical intervention) should **usually not be subjected to surveillance**.

While no decision should be taken on patient age alone, age is a core part of decision making for this patient group, where the expected timescale for serial surveillance before considering major prophylactic surgery is in the order of multiple years.

Older patients with a lower-risk aetiology (e.g. hypertension) and particularly those with only mild aortic dilatation, are less likely to ever meet surgical criteria in their lifetimes, due to low rate of progression. Clinical judgement regarding suitability should be applied before referring for surveillance. Where there is doubt, it may be appropriate to undertake shared decision making with the patient before initiating surveillance.

Conversely, some patients with very high-risk aetiologies (e.g. Marfans, Loews-Dietz and Turner syndromes, bicuspid aortic valve, associated aortopathies e.g. coarctation) may benefit from serial imaging despite a small absolute aortic size at presentation.

4 National criteria for genetic testing in suspected familial aortopathies are found at the National Genomic Test Directory (https://www.england.nhs.uk/wp-content/uploads/2018/08/rare-and-inherited-disease-eligibility-criteria-v2.pdf) page 16 and are quoted here:

- Thoracic aortic aneurysm or dissection with onset before age 50, OR
- Thoracic aortic aneurysm or dissection with onset before age 60 with a first degree relative with thoracic aortic aneurysm or dissection, OR
- Thoracic aortic aneurysm or dissection before age 60 with no classical cardiovascular risk factors, OR
- Thoracic aortic aneurysm or dissection before age 60 with features suggestive of aortopathy, e.g. arterial tortuosity, OR
- Clinical features suggestive of Loeys-Dietz syndrome, OR
- Features of Marfan syndrome giving a systemic Ghent score of ≥7, following assessment by a clinical geneticist or specialist with expertise in aortopathy, OR
- High clinical suspicion of a condition predisposing to aortic/arterial disease AND diagnostic testing for other conditions such as Ehlers Danlos syndrome (where indicated) has not identified a causative mutation
- Any deceased individual with a thoracic aortic aneurysm or dissection detected at autopsy meeting one of the above criteria and who have relatives who will benefit from cascade testing using a genetic diagnosis will be suitable for post-mortem genetic testing.

5 Both CT and MRI are appropriate modalities for accurate cross-sectional imaging of the entire aorta, and local service provision may influence the choice between either. MRI is often preferred, especially in younger patients, due to the absence of ionising radiation as well as the ability to concurrently assess ventricular and aortic valve pathology (e.g. severity of AR).

For some patients (e.g. where MRI is contraindicated, poorly tolerated or 3D SSFP non-contrast MRA has already been of poor diagnostic quality) CT should be the preferred modality. Where CT is used, it is strongly recommended to use ECG-gating for all aortic scans, as this reduces artefacts in the aortic root due to cardiac motion.

Whichever modality is selected, the ESC guidelines recommend that the same modality is used repeatedly for surveillance, using the same measurement methodology for consistency. If rapid progression is observed that might influence decision to intervene, this should be confirmed by a second modality.

6 Risk stratification (High / Moderate / Low Risk) is based on a combination of both clinical history and absolute aortic dimensions – see table above. At each scan, the patient may be recategorized to a different group depending on the findings (e.g. increased to a higher category if the aortic size is progressing towards the need for surgical intervention).

The suggested surveillance intervals of 5 years for Low Risk, 3 years for Moderate Risk and Annual surveillance for High Risk, are based on regional group consensus, and are consistent with all currently available international society guidelines as well as published papers from international expert centres. They are provided as a guideline only, and there will be specific cases, informed by discussion between the reporting Cardiac Imager and the referring Cardiologist, where these intervals are further tailored to specific patient needs. Rapid progression might merit even shorter intervals than one year.

7 Patients identified as being at particularly high risk, may benefit from Emergency Health Care Planning that identifies them as such to other healthcare professionals (e.g. GPs, Paramedics, Emergency Physicians), as well as highlighting symptoms that should prompt emergency assessment and imaging to exclude acute aortic syndromes.

8 During the long intervals proposed for aortic surveillance, some patients will develop other comorbidities, that then would preclude them from benefiting from surveillance and eventual prophylactic aortic surgery. Surveillance patients remain under the care of their referring Cardiologist who can assess whether it should be discontinued if no longer appropriate.

It would be appropriate for reporting Radiologists / Imaging Cardiologists to highlight patients who have remained stably in the Low/Moderate risk groups, and have now reached an age such that even “worst case” progression of aortopathy is unlikely to reach surgical criteria within the patient’s expected lifespan. The purpose of such a comment is to facilitate clinical review (which may have not been necessary during the intervening surveillance years) and consideration of whether further monitoring / imaging is still required.

9 Detailed advice on managing aortopathy in pregnancy is beyond the scope of this guideline. Many patients identified as such will have inherited aortopathies, and thus already be categorised as being high risk by our flow-chart. Clinicians should be aware that the proposed surveillance intervals **do not apply in pregnancy and puerperium,** as there is increased risk of progression in aneurysm size and of aortic dissection.

Patients planning to get pregnant or who become pregnant should have an up-to-date baseline MRI aorta pre-pregnancy, alongside referral to the specialist Cardio-Obstetric clinic. Further serial Echo/MRI scans may be recommended during pregnancy and the puerperium, guided by expert advice from the clinic. Some patients may be counselled not to become pregnant due to their risk of

aortic dissection. Surgical intervention on the thoracic aorta may be considered at smaller absolute aortic dimensions where pregnancy is being contemplated, so the "standard" cut-offs do not necessarily apply.

10 Referral for surgery is based on the ESC Aortic and Congenital Heart Disease guidelines. Once a patient has met criteria for referral for aortic surgery, this should be highlighted to the referring clinician who will usually arrange clinical review and onward referral to Cardiothoracic Surgery.

Further surveillance imaging is usually required post-operatively to assess for both surgical complications and progression of aortopathy in the rest of the aorta. The ESC recommends CT at 1 month to look for early complications (e.g. pseudoaneurysm), then at 6 months, 12 months and at further intervals depending on clinical risk.

We have elected to classify non-syndromic post-op thoracic aneurysm repairs which are stable at 1 year within the moderate risk group, but all post-aneurysm repairs with high-risk familial aortopathies, and those operated on for acute aortic dissection, as being high risk. We propose that they then undergo imaging surveillance in the same manner as pre-operatively.

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