What do Cardiologists Expect Regarding Echocardiogram in Aortopathies

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Abstract

The echocardiogram is an available and essential imaging tool for the evaluation of thoracic aortic diseases. This examination plays a significant role in the identification and diagnosis, as well as the follow-up of chronic cases. Its availability, portability, cost-effectiveness, and the absence of exposure to radiation or contrast make it an important tool for monitoring patients with known aortopathy, who usually require serial examinations for follow-up. Although it is inferior in the total and detailed characterization of the aorta in all its sections compared to aortic angiography or angioscopes, the echocardiogram allows for functional assessment of the aortic valve and valve apparatus. In acute aortopathies, it also enables the evaluation of complications and can be performed on hemodynamically unstable patients. This article will discuss the theoretical and practical aspects of the applicability of the echocardiogram in the context of aortopathies.

Echocardiogram in Aortopathies

The echocardiogram is a crucial imaging tool for assessing the thoracic aorta and related diseases, being widely used for outpatient evaluation.1,2 Given its broad availability in cardiological practice and the absence of screening recommendations for aortopathy in the general population, as well as the largest number of cases being asymptomatic and the high morbidity and mortality associated with the condition, this exam plays a vital role in identification and diagnosis. This is especially true for cases involving the proximal portions of the ascending thoracic aorta, which are better characterized using echocardiography. The exam’s availability, portability, cost-effectiveness, and lack of radiation or contrast exposure make it invaluable for monitoring patients with known aortopathy, who often require regular follow-up exams. While echocardiography is not as comprehensive as CT angiography or magnetic resonance angiography for aortic assessment, it provides functional evaluation of the aortic valve and apparatus. In acute aortopathies cases, it can also assess complications, being feasible even in hemodynamically unstable patients. The main uses of echocardiography in aortopathy settings are summarized in Central Illustration.

Echocardiogram in Chronic Aortopathies

During standard examinations, aortic diameters should be measured at the level of the aortic annulus, sinuses of Valsalva, sinutubular junction, and ascending aorta. For standardization, measurements should be taken from the external part of the anterior wall to the internal part of the posterior wall at the end of diastole in TTE. Conversely, in TEE, where the probe is positioned behind the heart, measurements should be taken from the external face of the posterior wall to the internal face of the anterior wall.3,4 The only exception is the measurement of the aortic annulus diameter, which, by convention, is taken from the inner surface of one wall to the other mid-systole, when the aortic valve leaflets are open. Measurements must be taken across the long axis parasternal window, with the aorta perpendicular to the probe for optimal image quality (Figure 2).

Normal aortic diameters vary based on the patient’s age, sex, body surface area, and height. Diameters above 40 mm are generally considered to be increased. However, for patients significantly taller or shorter than average, these values must be adjusted. Several methods have been suggested to adjust measurements, including the indexed aortic size,5 which is the aortic diameter divided by body surface area, applied to a nomogram that classifies aneurysm size as low, medium, or high risk. More recently, the ratio between the aortic size and height has been shown to be equivalent to the indexed aortic size.6

Aneurysms

True aortic aneurysms are defined as a dilation of the aortic segments exceeding 50% of the expected value for that segment or 50% larger than the adjacent healthy segment, with an intact...
Echocardiogram in aortopathies

**Chronic aortopathies**
- Assessment of aortic dimensions
- Aortic valve anatomical and functional assessment
- Assessment of focal diseases
- Clinical follow-up

**Acute aortopathies**
- Intimal flap identification
- Aortic valve anatomical and functional assessment
- Pericardial effusion assessment
- Segmental contractility assessment

Figure 1 – Aorta and its portions; rPA: Right pulmonary artery. Excerpt from Erbel et al.³
Aortic wall, meaning all layers of the wall surround the entire aorta. They should be described by their size, location, and morphology (fusiform or saccular). Fusiform aneurysms, the most common type, involve symmetrical dilation affecting the entire aortic circumference, while saccular aneurysms are characterized by localized dilation. False aneurysms, or pseudoaneurysms, result from bleeding within the aortic wall, leading to a periaortic hematoma that is contiguous with the aortic lumen. These can occur due to trauma, infection, or contained rupture of an aortic aneurysm, dissection, or penetrating ulcer.

Aortic root aneurysms, the portion best evaluated by TTE, are the most common, accounting for over 60% of all

<table>
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<tr>
<th>Table 1 – Echocardiographic views of the aorta</th>
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<tr>
<td><strong>TTE</strong></td>
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<td>Parasternal long and short axis</td>
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<td>Apical four chambers</td>
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<td>Suprasternal</td>
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<td>Subcostal</td>
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Excerpt from Evangelista et al.1 TTE: Transthoracic echocardiography; TEE: transesophageal echocardiography

Figure 2 – Measurement of the aortic ring mid-systole (1), from inner wall to inner wall, and sinuses of Valsalva (2), sinotubular junction (3), and ascending tubular aorta (4) during end diastole. LA: Left atrium; RPA: Right pulmonary artery. Excerpt from De Backer, Muñoz-Mosquera and Campens7 and Goldstein et al.2
Thoracic aortic aneurysms (TAAs), followed by descending aortic aneurysms (35%) and aortic arch aneurysms (less than 10%). They can be caused by hereditary diseases, congenital conditions, multifactorial degenerative issues, previous aortic dissection, inflammatory diseases, and infections. For patients with genetic diseases and a bicuspid aortic valve, which tend to affect the proximal portions of the TAA (except for Ehlers-Danlos syndrome type IV), TTE may be sufficient for screening. According to AHA recommendations, TTE should be performed on all patients with TAA dilation at the time of diagnosis to evaluate valve anatomy, valve function, and thoracic aorta diameters. TTE is also recommended for periodic follow-up in this group, alongside CT and MRI.

In patients with aortic root aneurysms accompanied by functional aortic insufficiency, several parameters should be assessed to determine the mechanism of insufficiency. This evaluation is crucial for deciding the appropriate surgical procedure, particularly whether valve-sparing strategies are viable, and should be conducted before any damage to the valve apparatus occurs. They speak in favor of functional aortic regurgitation with an anatomically normal aortic valve:

1. Dimensions of the ring, sinus of Valsalva, sinotubular junction and tubular aorta;
2. Leaflet coaptation height (tenting): maximum protodiastolic distance between the tip of the leaflets and the plane of the ring between 0.8 and 10 mm;
3. Sinotubular junction/aortic annulus ratio > 1.6 mm (mismatch between the dimensions of the junction and the annulus).

It has been demonstrated by La Canna et al. that the mismatch between junction and annulus is even more important than the isolated dimensions of the portions of the ascending TAA. Furthermore, factors such as significant calcification or wide fenestrations in the leaflets are parameters that indicate impairment of the valve apparatus.

**Focal Diseases**

Atherosclerotic plaques can be visualized using echocardiography as irregular, heterogeneous images or hyperreflective calcified foci attached to the endothelial layer of the vessel, often accumulating at the sinotubular junction and aortic arch. Plaques thicker than 5 mm, with mobile or prominent parts, ulcerations greater than 2 mm, and non-calcified plaques are associated with a higher risk of stroke, either spontaneously or during invasive procedures and open-heart surgery.

**Bicuspid Aortic Valve**

During the examination of a patient with a bicuspid aortic valve (congenital condition that affects 1 to 2% of the population), in addition to the routine assessment of parameters related to valve function and investigation of aortic insufficiency and stenosis, the TAA must be carefully evaluated. The diameters must be described as mentioned above, as the literature indicates that 20 to 84% have an associated TAA aneurysm. The descending aorta and arch must also be carefully examined in search of narrowing and acceleration of flow in the isthmus region, to rule out associated aortic coarctation.

**Echocardiography in Acute Aortic Syndromes**

Acute aortic syndromes (AAS) encompass a group of conditions with high morbidity and mortality, typically resulting from the loss of aortic wall integrity. These conditions are mainly formed by a triad: aortic dissection, aortic intramural hematoma, and penetrating atherosclerotic ulcer (figure 3).

Population studies suggest an incidence of AAS of 2.6–3.5 cases per 100,000 person-years, with 80% of these cases being aortic dissection, 15% intramural hematoma, and 5% penetrating aortic ulcer.

Given the high morbidity and mortality associated with AAS, it is crucial to achieve a quick and accurate diagnosis. Delay in recognizing these conditions is linked to increased mortality, with a 1% rise in the mortality rate for each hour of delay in definitive treatment (surgical correction), according to the literature.

Diagnosis of AAS can be multimodal, utilizing techniques like computed tomography (the most common method for diagnosing aortic dissection), magnetic resonance imaging, and echocardiography. Both TTE and TEE provide not only anatomical details of aortic involvement but also essential functional information for proper surgical planning. This includes assessing the mechanism and severity of aortic valve insufficiency, myocardial contractility changes that may suggest coronary artery involvement, and the presence of pericardial effusion, which can lead to hemodynamic compromise.

The sensitivity and specificity of echocardiography in diagnosing AAS vary depending on the method used (transthoracic or transesophageal) and the segment of the aorta examined. For TTE, sensitivity and specificity range from 77-80% and 93-96%, respectively, while diagnosing the ascending aorta dissection. However, for the descending aorta, the accuracy decreases, with sensitivity and specificity ranging from 31-55% and 60-83%, respectively. Nonetheless, as demonstrated by Evangelista et al., using contrast can significantly enhance the accuracy of TTE in evaluating both ascending and descending aortic dissections, potentially matching the sensitivity and specificity of the transesophageal method for diagnosing ascending aortic dissections. The TTE can reach sensitivity and specificity of 99% and 89%, respectively, in addition to a negative predictive value of 99%.

Table 2 provides a summary of the accuracy for the main imaging methods available for diagnosing SAA as well as its complications.

**Echocardiographic Evaluation in Aortic Dissection**

The diagnosis of a classic aortic dissection is made by demonstrating an intimal flap dividing the aorta into two lumens: a true and a false one. Often, the false lumen can be detected by color Doppler, but this may be not the case for thrombosis and retrograde dissection (Figure 4A and 4B). Identification of the true lumen is extremely important, as it helps understanding aortic involvement and its potential complications, as well as aids in surgical planning, if this is the indicated treatment option.

Some echocardiographic signs help in this differentiation. In general, the true lumen tends to have a smaller diameter and undergoes systolic expansion, while the false lumen has a larger diameter and undergoes systolic compression.

As previously mentioned, the TTE is especially useful for an adequate assessment of the root and proximal portion of the ascending aorta, while the TEE allows visualization of the entire
The thoracic aorta, except for the distal portion of the ascending aorta and the proximal portion of the aortic arch, the point of intimal laceration can be identified in approximately 78-100% of cases using the transesophageal method. It is usually located in the proximal portion of the ascending aorta in type A dissections and just after the origin of the left subclavian artery in type B dissections. Lacerations with diameters greater than 7-10 mm are typically easy to identify by 2D echocardiography (Figure 5). Secondary communications between the two lumens can be observed by echocardiography, and these must be differentiated from the main laceration points. Some anatomical studies suggest that these communications, often seen in the descending aorta on color Doppler, represent the origin of intercostal or visceral arteries and generally have a diameter smaller than 3 mm.

Table 2 – Comparison of the Diagnostic Accuracy of the Main Imaging Modalities in AASs

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<tr>
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<td>Ascending aortic dissection</td>
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<td>Aortic arch dissection</td>
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<td>Descending aortic dissection</td>
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<td>Intramural hematoma</td>
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<td>Penetrating aortic ulcer</td>
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<td>Aortic valve morphology and function</td>
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<td>Dynamic flap view</td>
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<td>Dimensions of the aortic lumen</td>
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<td>False lumen thrombosis</td>
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<td>Ventricular function</td>
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<td>Hemodynamic assessment</td>
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<td>Assessment of coronary involvement</td>
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<td>Assessment of involvement of aortic branches</td>
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Adapted from Evangelista et al.13 AAS: acute aortic syndromes.
Among the possible complications of aortic dissections, pericardial effusion (Figure 6) is notable, present in up to 40% of cases of type A dissections and accompanied by tamponade in 12% of such cases.1,7 In this context, echocardiography is the best imaging technique to evaluate cardiac tamponade. Pericardial effusion is not always secondary to rupture of the false lumen into the pericardium; it may also be caused by irritation of the adventitia due to an aortic hematoma.12 Periaortic hematoma is another reported complication associated with increased mortality and is best diagnosed by computed tomography.1,12

Aortic insufficiency (Figure 7) is also a frequent complication, occurring in approximately 40-76% of cases of type A dissection.1,12 Besides assessing the severity of regurgitation, echocardiography helps understand the mechanism of aortic insufficiency and assists surgeons in choosing the best surgical technique, whether it involves preservation or replacement of the aortic valve.3

Coronary involvement due to aortic dissection can occur in around 10-15% of cases, with the right coronary artery being most affected. Alteration in ventricular segmental contractility helps in the diagnosis of this complication.1 The transesophageal method allows for the evaluation of the proximal portion of the coronary arteries and verification of their origin from the true or false lumen, as well as the presence of dissection.1

However, echocardiography has limitations in evaluating aortic dissection, mainly due to artifacts that can create a false appearance of a dissection flap. The aortic wall can cause a reverberation artifact within the screening sector, simulating a dissection flap (Figure 8). Color Doppler can aid in differentiation, as a true flap will show two distinct areas of color flow, one for the true lumen and another for the false one. Besides, M-mode can also help by demonstrating independent movement of the true flap in relation to the aortic wall, unlike the false flap, which will present identical movement.14

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**Figure 4** – Intimal flap dividing the aortic root into two lumens, one true and the other false (a). The use of color Doppler can help identify the true lumen, as in the case of image (b) of a patient with abdominal aortic dissection. Adapted from Evangelista et al.1 LV: left ventricle.
Figure 5 – View of the descending aorta in the short axis using the transesophageal method. To the left, the true lumen (arrowhead) and the false lumen (asterisk), separated by the flap (arrow), can be identified. To the right, the laceration point is confirmed by the Color Doppler, which indicate the flow from the true lumen to the false one. Excerpt from Eissa et al.16

Figure 6 – Image obtained through the transthoracic method, using the subcostal window. A flap in the aortic root (white arrow) can be seen, as well pericardial effusion causing compression in the right ventricle (black arrows). Excerpt from Evangelista et al.12 RV: right ventricle; Ao: aorta; LV: left ventricle.
Echocardiographic Evaluation of Intramural Aortic Hematoma

Intramural hematoma is characterized by a thickening of the aortic wall with a concentric growth pattern, typically preserving the vessel’s lumen. Diagnosing this condition can be challenging, as initial imaging tests can be normal in up to 12% of patients. While the transthoracic method is often not suitable for diagnosing this condition due to its low sensitivity, the transesophageal method offers better accuracy, though still inferior to computed tomography and magnetic resonance imaging. The diagnostic criteria for intramural hematoma using the transesophageal method is defined as a thickening of the aortic wall > 5 mm, and a cutoff point of > 7 mm, which can be used for patients with severe atherosclerosis (Figure 9).

Echocardiographic Evaluation of Penetrating Atherosclerotic Aortic Ulcer

Penetrating atherosclerotic ulcer is defined as an ulceration of the atherosclerotic plaque that penetrates the inner layer of the aortic media. This condition can coexist with intramural hematoma, may be multiple and most commonly affects elderly individuals. Although it can be located in any aortic segment, it is more commonly found in the middle and distal portion of the descending thoracic aorta. Computed tomography is superior in identifying this condition, but the aortic ulcer can still be visualized using the transesophageal method, typically appearing as a crater with irregular edges associated with multiple intimal irregularities. Color Doppler can be useful to demonstrate turbulent flow within and at the ulcer entrance (Figure 10).

Figure 7 – Images obtained by TEE. To the left, the intimal flap near the valve plane can be observed, and to the right, the aortic valve insufficiency caused by the dissection. Adapted from D’Aloia et al.
Figure 8 – Image obtained by the transthoracic method, using parasternal long axis window. A reverberation artifact is noted in the ascending aorta (red arrow) due to calcification at the sinotubular junction, which may simulate a dissection flap. Adapted from Bertrand et al. 19

Figure 9 – Images obtained by the transesophageal method can show thickening of the aortic wall (arrows). Adapted from Evangelista et al. 13
Conclusion

Echocardiography plays an essential role in the multimodal assessment of aortopathies. Like other imaging techniques, this method has both strengths and limitations. The clinician’s understanding of the method is crucial, as its precise application can provide significant answers and complement information from other techniques. This improves patient care within the complexity of acute and chronic thoracic aorta conditions.

Author Contributions

Writing of the manuscript and critical revision of the manuscript for intellectual content: Martinhago GA, Andrade AR, Correia VM, Fernandes F, Santiago JAD, Dias RR, Madrini Junior V.

Potential Conflict of Interest

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Study Association

This study is not associated with any thesis or dissertation work.

Ethics Approval and Consent to Participate

This article does not contain any studies with human participants or animals performed by any of the authors.

References


Figure 10 – Images obtained by the transesophageal method showing atherosclerotic ulcers penetrating the aortic wall. Adapted from Evangelista et al.13


