Aortic Arch Vessel Disease and Rationale for Echocardiographic Screening

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Atherosclerosis of the proximal branches of the aortic arch has compelling clinical implications that warrant the application of direct noninvasive detection of the disease. The prevalence of aortic arch vessel disease in an aging and at-risk community and clinical population has been underreported and undertreated despite an associated increase of all-cause and cardiovascular mortality. Intrathoracic duplex imaging has been validated as an accurate noninvasive tool to detect, characterize, and follow native aortic arch vessel disease and its sequelae and correction. Such duplex techniques are easily integrated into routine echocardiography with focused training and minimal time investment in the examination. A paucity of available resources exists across disciplines regarding ultrasonographic investigation of these supra-aortic trunk vessels, including textbooks, journal articles, seminars, and manuals. This review has been compiled to familiarize physicians and sonographers with the relevant anatomy, pathophysiology, treatment, and diagnostic duplex surveillance of aortic arch vessel disease. Illustrative cases along with clinical rationale are discussed with the intent to facilitate the integration of arch vessel duplex imaging into the scope and practice of echocardiography. (J Am Soc Echocardiogr 2013;26:114-25.)

Keywords: Echocardiography, Aortic arch, Intrathoracic duplex sonography, Aortic arch vessel disease, Subclavian steal syndrome

Although the suprasternal notch is a standard echocardiographic window for the assessment of aortic arch and aortic valve pathologies, its potential for evaluation of the proximal aortic arch vessels has been largely ignored. Patients undergoing extracranial cerebrovascular and upper extremity arterial examinations for cerebral ischemia symptoms form an indication-appropriate subset of echocardiogram referrals.[1] However, intrathoracic duplex imaging of the proximal subclavian and carotid trunks is not typically performed with these vascular modalities. Thus, these aortic arch vessels are a frequently overlooked cardiovascular territory.[2] Invasive digital subtraction angiographic studies along with emerging applications of magnetic resonance angiography and computed tomographic angiography (CTA) are used for definitive anatomic characterization of arch vessel lesions, particularly when endovascular and surgical interventions are entertained. Duplex imaging of aortic arch vessel disease (AAVD) has proven accuracy in characterizing lesion significance and, when integrated with echocardiographic protocols, has the potential to improve screening and detection. Morphologic and hemodynamic information found in this approach is a valuable complement to noninvasive vascular and angiographic imaging in guiding clinical decision making. Additionally, asymptomatic and noncritical lesions detected by such echocardiographic techniques have sound therapeutic and clinical implications that warrant its use as a routine screening tool for AAVD. An anatomic overview of the aortic arch and its proximal branch vessels will precede a description of how to examine these targets optimally, followed by illustrative cases and discussion designed to demonstrate clinically relevant pathology and applications.

ARCH VESSEL ANATOMY

Several normal anatomic variations and anomalous vessel branching patterns radiating from the outer curvature of the aortic arch have been described. The standard arch configuration shows a shared brachiocephalic trunk, called the innominate artery (IA), which bifurcates superiorly, giving rise to the right common carotid artery (RCCA) and right subclavian artery (RSA). The left common carotid artery (LCCA) and left subclavian artery (LSA), in downstream sequence, have separate origins (Figure 1A).[3] The diameter of these supra-aortic trunk vessels ranges from 5 to 9 mm, averaging 7.2 $\pm$ 0.6 mm, determined angiographically.[4] The so-called bovine arch variation is apparent when the LCCA merges proximally off of a single trunk in common with the IA. In this instance, occurring overall in 13% of the general population (25% of African Americans, 8% of whites), there are a total of two brachiocephalic origins (Figure 1C).[5] The bovine namesake is actually a misnomer, as the true bovine pattern found in cattle manifests a single brachiocephalic trunk from the arch. Yet a third anatomic variation occurs when the LCCA has a more distal, distinct origin from the IA trunk and is seen in about 9% of the population

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Abbreviations

AAVD = Aortic arch vessel disease

CABG = Coronary artery bypass grafting

CAD = Coronary artery disease

CSSS = Coronary-subclavian steal syndrome

CTA = Computed tomographic angiography

DSA = Digital subtraction angiography

IA = Innominate artery

IAPD = Interarm blood pressure difference

IMA = Internal mammary artery

LCCA = Left common carotid artery

LSA = Left subclavian artery

PAD = Peripheral arterial disease

RCCA = Right common carotid artery

RSA = Right subclavian artery

SAS = Subclavian artery stenosis

(10% of African American, 5% of whites) (Figure 1B). Additionally, two rare anatomic variants occur, one with a common left brachiocephalic trunk (LCCA and LSA) and another as a separate vertebral artery originating from the arch, seen in 1% and 0.5%, respectively (Figure 1D).[6] There are multiple rare arch vessel branching anomalies that can be classified according to the position and caliber of the aorta and number of abnormalities. The most common aberration is the left aortic arch that gives rise to an anomalous RSA (0.4%–2%). A bicarotid trunk arising from the aorta is also found in association with an anomalous origin of the RSA as it branches from a common subclavian trunk at a more distal arch position. The right aortic arch with an aberrant LSA and double aortic arch are both associated with vascular rings that can encircle the trachea and esophagus, causing symptomatic impingement of those structures. The right aortic arch with mirror image branching, and left-sided or right-sided cervical arches associated with vessel branching variations, are seen among other rare congenital malformations.[7,8] Patients with clinically significant arch anomalies are imaged best noninvasively using magnetic resonance angiography and CTA. Familiarity with these aortic arch and vessel attributes is fundamental to the acquisition and interpretation of duplex images demonstrating arch vessel disease.

EXAMINATION TECHNIQUE

Duplex surveillance of the intrathoracic supra-aortic trunk vessels may be accomplished as an adjunct to the standard echocardiographic examination of the aortic arch. The supine patient is instructed to slightly hyperextend the neck and, if necessary, turn the head to the right, facilitating placement of the ultrasound probe in the suprasternal notch. The long axis of the aorta is displayed using a 2-MHz to 5-MHz multifrequency phased-array transducer with system settings to maximize resolution. High-resolution harmonic imaging is beneficial, as is sacrificing frame rate in favor of improved image detail, because the temporal resolution normally required for dynamic cardiac motion is not an issue with these structures. Usually, the arch vessels are readily apparent on grayscale imaging and can be visualized in their longitudinal orientation either simultaneously with the arch or with minimal manipulation (Figure 2A). Qualitative color flow Doppler imaging is routinely used to differentiate and locate the arch vessels and discern laminar versus disturbed flow patterns (Figure 2B). The color Doppler scale should be set to 40 to 60 cm/sec and optimized by adjusting the color box size, image depth, sector size, and gain.[9] More extensive quantitative Doppler sampling is reserved for absent or disturbed flow signals.

Careful inspection of the arch vessels typically begins with the LSA and continues in a retrograde and medially directed fashion to include the LCCA, followed by the IA and its branches. Color Doppler optimization will help identify the LSA takeoff signal, whose flow stream tends to be somewhat perpendicular to the incident beam. The LSA is also deeper and prone to reverberation image artifact, whereby one or two weaker "ghost" color flow Doppler signals may appear posteriorly.[10] When image quality is marginal, this artifact presents an obstacle to confirming a variant separate origin of the left vertebral artery directly from the arch. Highly suboptimal arch images, although not uncommon, do not preclude inspection of the arch vessels. In such cases, a strategy of finding the more distal vessels and following them back to their arch origins, using complementary color flow and spectral Doppler, is frequently successful. For example, by sliding the probe over the left sternocleidomastoid muscle or into the left supraclavicular fossa, the distal portion of the LSA may be identified and traced back to the arch, allowing inspection of its proximal takeoff. The LCCA has a distinct curvilinear course and is typically imaged along its proximal trunk without difficulty. Its ease of imaging and distinguishing features can be used as a helpful landmark to locate the other arch vessels in technically challenging examinations. Slight counterclockwise probe rotation while migrating superiorly will permit visualization of the IA, frequently laying out its RCCA and RSA bifurcation, whereby the proximal and quite superficial portions of these trunks can be evaluated (Figure 3). When unable to demonstrate the IA bifurcation, the RSA and RCCA should be located and examined individually. Following the natural cephalad course of the IA from the usual echocardiographic arch image will allow swift screening and visualization of its branching with minimal probe manipulation. Although the display of RCCA and RSA branching is anatomically reversed, retaining standard arch orientation and continuity is essential particularly when technically difficult examinations require reliance on color flow mapping to identify relative arch vessel positions. Electronic left-to-right image inversion of the bifurcation of the IA trunk can be used, if desired, to reorient the vessels to anatomic and angiographic views, especially when demonstrating lesions. Electronically inverted images should be clearly annotated as such on the image to avoid confusion. The shallow branches of the IA and more superficial courses of the other arch vessels, including the vertebral artery origins are best imaged with high-frequency linear-array transducers. This is normally integrated within the extracranial carotid survey, which may also include provocative arm exercise or reactive hyperemia to assess vascular steal.

Careful attention to these arch vessel imaging techniques permits efficient and accurate qualitative confirmation of normal anatomy and laminar color Doppler flow signals. A study published in 1984 by Ackerstaff *et al.*[11] identified 112 of 118 (95%) of angiographically normal subclavian arteries. Another report on 20 patients with Takayasu's arteritis found that duplex imaging, validated by angiography, correctly distinguished normal vessels in 12 of 15 IAs (80%), 12 of 13 LCCAs (92%), and all subclavian arteries.[12] In our experience, normal arch vessel anatomy and flow are confirmed by adding 30 to 60 sec to the echocardiographic aortic arch examination.

When color flow Doppler turbulence is apparent, best adjustment of the real-time image and quantitative Doppler interrogation is performed. Split-screen comparative display techniques with and without color flow Doppler are often helpful by providing anatomic context with the arch and proximal vessel trunks. After image depth and quality