American College of Radiology
ACR Appropriateness Criteria®

Clinical Condition: **Imaging in the Diagnosis of Thoracic Outlet Syndrome**

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	8		☢
MRA chest without and with IV contrast	8		O
CTA chest with IV contrast	7		☢☢☢
MRI chest without IV contrast	7		O
US duplex Doppler subclavian artery and vein	6		O
Digital subtraction angiography upper extremity	5		☢
CT chest without IV contrast	3		☢☢☢
MRA chest without IV contrast	2		O
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

IMAGING IN THE DIAGNOSIS OF THORACIC OUTLET SYNDROME

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Summary of Literature Review

Introduction/Background

Thoracic outlet syndrome (TOS) is a clinical entity characterized by compression of the neurovascular bundle of the upper limb as it passes from the upper thoracic aperture to the axilla. Although thrombosis of the axillosubclavian vein was first reported by Paget in 1875 and Von Schroetter in 1884, and was coined “Paget-Schroetter syndrome” by Hughes in 1949, the term “Thoracic Outlet Syndrome” was coined in the 1950s to reflect the fact that TOS has many variants, ranging from vascular involvement of the subclavian artery (SCA) or vein (SCV) to the more common neurogenic form with compression of the brachial plexus.

TOS most commonly occurs in females (4:1) between the ages of 20 and 40 [1]. The site of compression occurs either at the interscalene triangle, the costoclavicular space, or the retropectoralis minor space [2,3]. However, as the thoracic outlet is defined anatomically as the interscalene space, and compression is most common at this site, it has been proposed to more precisely categorize patients into TOS, costoclavicular and pectoralis minor subtypes [4,5]. Both congenital and acquired etiologies may play a role, including bony issues such as first rib abnormalities, cervical ribs and bony tubercles, and soft-tissue anomalies such as fibrous bands, cervical muscle hypertrophy, or postural problems such as drooping or sagging of the shoulders [6-8].

Symptomatology will vary depending on the site of compression and the nature of the compressed structure; clinically, patients can be divided into neurogenic, arterial, venous, mixed (neurogenic and vascular), and nonspecific subtypes [5]. Neurogenic symptoms are most common, accounting for greater than 90% of all TOS cases [9]. In neurogenic TOS (nTOS), compression of the peripheral nerves of the brachial plexus gives rise to pain, paresthesia, dysesthesia, or weakness in the upper limb. Vascular involvement may present with swelling, edema, skin changes, or upper limb weakness and fatigue on exercise. This diverse presentation makes the true incidence of TOS unknown, although it has been estimated as 5 per 100,000 [10].

Evaluation of potential TOS comprises a careful examination to discern any arterial, venous, or neurologic compromise. To this end, evaluation of peripheral pulses and pressures is followed by neurologic testing using compressive or provocative tests such as Tinel sign, Adson test, Roos test, Wright test, and the hyperabduction maneuver. Radiography (to evaluate for cervical ribs or first rib bony spurs) and electrodiagnostic tests such as brachial plexus neurography are typically performed [11] prior to further imaging.

The goal of further imaging is to confirm the diagnosis of TOS, exclude mimics such as cervical spondylosis or shoulder joint or lung apex pathology, allow accurate classification into nTOS versus venous (Paget-Schroetter) versus arterial TOS, and guide treatment selection to minimize morbidity and mortality. Complications of vascular involvement include arterial compression leading to limb ischemia, or embolic phenomena from arterial thrombosis causing stroke or digital ischemia [12]. With venous involvement, deep venous thrombosis (DVT) can lead to postphlebitic limb, with the feared complications of phelgmasia cerulea dolens and venous gangrene in

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severe cases. The risk of pulmonary embolism (PE) in upper-limb DVT is controversial, with some authors stating that the risk of incidence of PE attributable to previously documented upper extremity DVT is very small (1%) regardless of anticoagulant therapy [13], whereas other authors postulate a risk of PE of up to 9% [14].

Treatment of TOS is based on the causing etiology, the symptom complex, and the presence of complications at the time of diagnosis. In venous TOS the role of venous thrombolysis prior to surgical decompression has gained widespread favor [15-17], although the timing of subsequent surgery remains debatable [6,18]. Although surgical first rib resection, via either an anterior or transaxillary approach, remains the gold standard for decompression of TOS [19], minimally invasive methods of treatment of nTOS, including scalene blocks or botulinum toxin injection with either ultrasound (US) [11] or computed tomography (CT) guidance [20], are gaining favor.

Overview of Imaging Modalities

The goal of imaging, regardless of modality, is to localize the site of compression, the compressing structure, and the compressed organ or vessel. By convention, abduction of the upper limb has been shown to be relevant in diagnosing TOS and is thus chosen as the postural maneuver of choice for cross-sectional imaging [21-23]. In the abduction of the upper limb, narrowing of the subclavian vessel is considered significant if the percentage change of the vessel's diameter between the neutral and the abducted positions is 30% or greater for the SCA and 50% or greater for the SCV [2].

Digital Subtraction Angiography

Although historically direct arteriography or venography would have been considered the gold standard for evaluation of extrinsic compression of the subclavian vessels, the lack of visualization of the impinging structure and nonvisualization of neurologic structures means digital subtraction angiography and venography is almost exclusively reserved for intraprocedural interventional guidance.

Chest Radiography

Chest radiographs are frequently used as an initial imaging modality in suspected TOS due to the ease of access, safety profile, low cost, and the ability to evaluate several of the osseous abnormalities associated with TOS. These include first rib anomalies [7], cervical ribs [24], congenital osseous malformations [25,26], and focal bone lesions [27]. Soft-tissue lesions, such as lung neoplasms [28], may also be evaluated, although the negative predictive value of chest radiography is debatable.

Ultrasound

US is widely used as a cost-effective, safe, and quick imaging modality in the initial evaluation of patients with either arterial or venous pathology throughout the body. Real-time duplex US is noninvasive and can be easily performed during dynamic maneuvers.

The technique involves B-mode US and Doppler study of the subclavian vessels, typically performed at rest (neutral position) and with provocative maneuvers such as Adson, Eden, and 90° Wright tests. These tests were considered positive if they produced flow acceleration followed by turbulence and, finally, by an arrest in signal propagation [29,30]. Evaluation of the cross-sectional area of the costocervical space may also be performed [31]. For venous TOS, US has a longstanding and well-documented role in the diagnosis of upper extremity DVT [18,32].

However, although the main advantage of US is the ability to directly compare between provocatively induced symptoms and concurrent direct vessel visualization, there is debate in the literature as to the significance of imaging findings, particularly with respect to maneuvers to minimize the thoracic outlet and associated spaces as described above [29-31,33]. Moreover, although visualization of the vessels is a strength of US, sonographic diagnosis of compressive effects upon the brachial plexus is a challenge [34], and symptoms of TOS may unmask a deeper regional pathology such as Pancoast tumor or cervical spondylopathy, requiring further imaging.

Computed Tomography Angiography

Contrast-enhanced CT evaluation of TOS is typically performed as a 2-step procedure in which initial "neutral" images are obtained from elbow to aortic arch with the arms adducted to the side, followed by abduction and repeat imaging in an effort to reproduce the neurovascular compression seen on provocative maneuvers. Some centers add the additional step of placing the contralateral arm in abduction (with the symptomatic ipsilateral arm in the neutral position) in order to minimize streak artifact [35]. Scan acquisition is typically performed with a contralateral antecubital injection of contrast material, with either an empiric scan delay of 15–20 seconds or bolus tracking over the ascending aorta [36,37].

Multiple studies have demonstrated the utility of CT in evaluation of the upper-limb arteries and veins; however, reliance on axial slices alone can lead to misrepresentation of the degree of any stenosis, with one study showing underestimation of stenosis found in 43% of transverse CT scans but only 10% of sagittal reformations. Overestimation of stenosis was also more frequent on surface displays with 3-D shading (16%) than on volume-rendered images (7%) [36], advancing the case for evaluation of these studies on vascular workstations. Beyond the vessels themselves, CT allows quantification of the change in costoclavicular or interscalene spaces with provocative maneuvers [36,37], the presence of bony abnormalities [7,38], or superior sulcus pathology [39].

The efficacy of CT in evaluation of TOS depends greatly on the classification. For arterial compression, there is evidence of good correlation of CT findings with operative findings and results of decompression [40]. However, venous findings are less predictive due to the prevalence of compression of the vein frequently seen in all compartments of the thoracic outlet after arm abduction [41,42]. CT for the evaluation of nTOS is limited by the lack of contrast resolution of neural structures, although evaluation of the space sizes gives secondary indicators that may aid in diagnosis [37].

Magnetic Resonance Imaging/Magnetic Resonance Angiography

Magnetic resonance imaging (MRI) is now a widely available and utilized modality for reliable, reproducible, noninvasive, and nonionizing evaluation of the vasculature, nervous system, and soft tissues [43]. MRI has inherent advantages over US in its ability to delineate extravascular anatomy, particularly in anatomic sites with poor sonographic windows, and it has advantages over CT in its characterization and differentiation of soft tissues. MRI does, however, have contraindications and is not recommended in certain patients, such as the very obese, claustrophobic, or those with MRI unsafe devices.

MRI has been shown to accurately demonstrate upper-limb arterial and venous thrombus, using both contrast-enhanced and noncontrast sequences [44,45]. Typically MRI for TOS is performed with high-resolution T1-weighted and T2-weighted sequences in sagittal and axial planes to delineate anatomy and evaluate cervical radiculopathy, the brachial plexus, muscular attachments, and sites of compression [46,47]. Evaluation of the vasculature is then performed in both neutral and arms-abducted positions. Noncontrast-enhanced MRI can be sufficient to diagnose nTOS. This is the most common variant of TOS and may be the sole manifestation of the syndrome or occur in conjunction with vascular obstruction [41,46]. In patients with nTOS, sagittal T1-weighted imaging performed with patient's arms in abduction typically demonstrates effacement of fat adjacent to the brachial plexus roots, trunks, or cords within the interscalene triangle or costoclavicular space. T1-weighted imaging performed in sagittal and axial planes can also demonstrate causative lesions of nTOS, including cervical ribs, congenital fibromuscular anomalies, and muscular hypertrophy (eg, subclavius muscle). Imaging with turbo-spin echo T2-weighted or short tau inversion recovery sequences can be useful in cases where spinal cord lesions or primary disorders of the brachial plexus (eg, brachial plexitis) are considered as alternative diagnoses to nTOS [43].

For noncontrast time-of-flight (TOF) imaging, a saturation band can then be applied medial to the imaging slice for the SCV, lateral for the artery, and since no intravenous contrast is required, a potential advantage of TOF imaging is the ability to repeat acquisitions in different stress positions without venous contamination. Among the limitations of noncontrast imaging are intraluminal filling defects related to flow or in-plane saturation effects. Hence the majority of MR angiography/MR venography (MRA/MRV) for TOS is performed with contrast-enhanced sequences. A recently published study described a protocol [12] on either 1.5-T or 3.0-T MRI scanners whereby breath-hold arterial and venous-phase contrast-enhanced 3-D MRA and MRV/equilibrium-phase images were obtained with a 3-D gradient-echo pulse sequence with fat suppression. Images were obtained in both the arms-abducted and neutral positions. Extracellular contrast agents were used, although blood pool agents, although perhaps limiting pure angiographic rendering in both positions, may facilitate a high-quality venous and arterial study. Use of open scanners for evaluation of TOS has also been reported [23].

Use of either contrast-enhanced or noncontrast imaging depends on factors such as patient renal and respiratory function and expertise and experience of the radiologist. However, although both forms of MRA may demonstrate TOS with significant arterial impingement, in comparison with TOF sequences contrast-enhanced MRA generally offers extensive vessel coverage, is less prone to artifact, and more frequently demonstrates the underlying cause of TOS when studies are reformatted [48]. Several studies have demonstrated excellent utility of contrast-enhanced MRA in showing a significant difference in MRA/MRV findings between the neutral and provocative positions [12,48,49]. Of note, however, care is needed as compression of vessels, particularly the vein, can be seen in up to 47% of normal patients with dynamic maneuvers [21], so clinical evaluation and correlation is vital.

Summary

- TOS is characterized by compression of the neurovascular bundle as it passes from the upper thorax to the axilla. There are arterial, venous, neurogenic, and forms.
- It may be congenital or acquired and may be secondary to bony issues such as first rib abnormalities, cervical ribs and bony tubercles, or soft-tissue anomalies such as fibrous bands or cervical muscle hypertrophy.
- The goal of further imaging is to confirm the diagnosis of TOS, exclude mimics such as cervical spondylosis or shoulder joint or lung apex pathology, allow accurate classification into neurogenic TOS versus venous (Paget-Schroetter) versus arterial TOS, and guide treatment selection to minimize morbidity and mortality.
- Abduction of the upper limb has been shown to be relevant in diagnosing TOS and thus is the postural maneuver of choice for cross-sectional imaging.
- Digital subtraction angiography, US, CTA, and MRA may allow evaluation of vascular structures and the secondary effects of compression, whereas CT and MR allow identification and evaluation of surrounding neurologic, soft-tissue, and bony structures.

Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, both because of organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared to those specified for adults (see Table below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® Radiation Dose Assessment Introduction document.

Relative Radiation Level Designations		
Relative Radiation Level*	**Adult Effective Dose Estimate Range**	**Pediatric Effective Dose Estimate Range**
O	0 mSv	0 mSv
☢	<0.1 mSv	<0.03 mSv
☢☢	0.1-1 mSv	0.03-0.3 mSv
☢☢☢	1-10 mSv	0.3-3 mSv
☢☢☢☢	10-30 mSv	3-10 mSv
☢☢☢☢☢	30-100 mSv	10-30 mSv

*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as "Varies".

Supporting Documents

For additional information on the Appropriateness Criteria methodology and other supporting documents go to www.acr.org/ac.

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