Review Article on Compressive Vascular Syndromes

Arterial thoracic outlet syndrome

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Abstract: Thoracic outlet syndrome (TOS) is used to describe the constellation of symptoms arising from neurovascular compression of the thoracic outlet. The structures passing through the thoracic outlet include the subclavian artery, subclavian vein and trunks of the brachial plexus. Patients may experience symptoms related to compression of any one or various combinations of these structures. Arterial pathology as the cause of TOS is rare, though repetitive overhead arm motion, such as seen in athletes, is a risk factor for developing arterial TOS (aTOS). Symptoms include chronic findings, such as pallor, arm claudication or cool arm. Currently diagnosis of aTOS is made using clinical and imaging parameters which include focused history and physical including provocative maneuvers and imaging follow-up ranging from angiography to MRI. Occasionally, acute thrombosis can result in limb threatening ischemia requiring emergent catheter directed thrombolysis. Outside of acute limb ischemia, management of aTOS is variable, however typically begins with conservative measures such as physical therapy. In patients who do not respond or progress on conservative management, surgical decompression may be performed. Open or endovascular treatment of subclavian artery pathology may be necessary for recalcitrant cases. In this article, the aim is to review the elements involving diagnosis and management of aTOS.

Keywords: Thoracic outlet syndrome (TOS), arterial thoracic outlet syndrome (aTOS), compression syndromes

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Introduction

Thoracic outlet syndrome (TOS) is commonly used to describe a constellation of symptoms such as arm pain, paresthesia, and weakness, thought to result from compression of the neurovascular bundle at the thoracic outlet (1). Approximately 2% of the North American population is affected by symptoms of TOS and is more common in women (2). TOS is typically categorized into neurogenic, venous, or arterial in nature. Neurogenic TOS (nTOS) is by far the most common comprising of approximately 95% of known cases whereas vascular causes are much less common with 4% being attributed to venous, and 1% resulting from an arterial etiology (3). The focus of this review will be to explore the anatomy, pathophysiology, diagnosis, management, and treatment of arterial TOS (aTOS).

Anatomy

The thoracic outlet is comprised of the space between the supraclavicular fossa and axilla. Particular attention should be drawn to the interscalene triangle, costoclavicular space, and subcoracoid space. The interscalene triangle is created by the anterior scalene (anterior border), middle scalene (posterior border), and first rib (inferior border) and is what the subclavian artery passes though along with the brachial plexus. The costoclavicular space is confined by the subclavius muscle (anterior border), anterior scalene (posterior border), first rib (inferior border) and clavicle

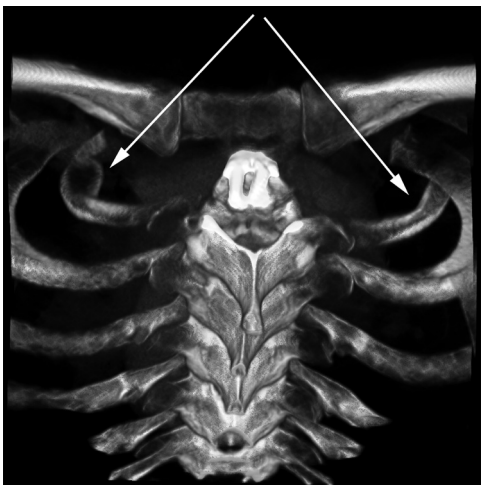

Figure 1 3D volume rendering demonstrates bilateral cervical ribs (arrows).

(superior border) and this is where the subclavian vein converges with the artery and brachial plexus to travel through this space. The most lateral space is the subcoracoid space bound by pectoralis minor (anterior border), 2–4 ribs (posterior border) and coracoid (superior border) through which the axillary artery, vein, and brachial plexus pass through. Alterations at these spaces, whether physiologic or pathologic, result in the variations of TOS that are seen (4).

Pathophysiology

There are a myriad of causes resulting in TOS including anatomic variations, blunt force trauma, repetitive microtrauma, muscle hypertrophy, and iatrogenic sources. During extreme shoulder abduction, the thoracic outlet is physiologically narrowed. As such, those who perform repetitive overhead arm movements such as swimming or throwing, are at risk of developing aTOS. Nearly half of patients presenting with aTOS have cervical ribs and around a third have soft tissue anomalies (3). Blunt force trauma can result in direct vascular injury or mass effect from displacement of bony structures and resulting hematomas account for nearly 5% of the cases of aTOS. aTOS can also result from any repetitive motions that spawn inflammation, microhemorrhage/fibrosis. Iatrogenic causes are possible, particularly in the setting of orthopedic interventions (5).

Presentation and diagnosis

The clinical diagnosis of thoracic outlet syndrome is challenging and there are no established criteria for diagnosis. Physicians ranging from primary care to specialists are involved in the care of patients with TOS. Presentation of aTOS can be partitioned into acute thrombosis, chronic stenosis, non-thrombotic ischemia, distal embolization or total occlusion (6). Arm claudication, coldness, and pallor may point to chronic stenosis or occlusion. aTOS can also occasionally manifest as non-limb related complaints such as a posterior circulation stroke (7). Clinical evaluation can include focused history of prior trauma, athletic history, exacerbating factors as well as provocative maneuvers such as the 90-degree abduction external rotation test (ROOS) while evaluating distal pulses (8). Unfortunately, patients presenting with vascular TOS have worse functional impairment compared to those with nonvascular symptoms (7).

Imaging workup includes radiographs of the neck to evaluate for supernumerary or ectopic ribs (*Figure 1*), CT, MRI/MRA, Doppler ultrasounds of the axilla as well as conventional angiography. Causes of aTOS can be divided into bony vs non-bony etiologies. Bony causes can be congenital such as a cervical rib (*Figure 2*) or bone tumors or related to trauma such as clavicular fracture (*Figure 3*). Nonbony causes include scalene hypertrophy, post-surgical scarring, or intrinsic arterial disease. Frequently, patients will present with a mix of symptoms with concurrent nTOS, venous (vTOS), and aTOS occurring simultaneously (*Figure 4*). It is therefore possible for a patient to present with neurologic symptoms such as paresthesias (nTOS) and venous symptoms such as deep venous thrombosis (vTOS) and be found to have arterial narrowing or occlusion (*Figure 5*).

Radiography of the chest is insufficient to diagnose aTOS however may be used in conjunction with other imaging methods to identify rib anomalies, osseous malformations or other bone lesions that may cause thoracic outlet impingement. Ultrasound Duplex Doppler may also be performed to identify abnormalities of the subclavian artery, and is particularly useful in showing dynamic changes with arm movement, however is limited in identifying specific etiologies of aTOS. CT chest may demonstrate anatomical narrowing of the thoracic outlet and can also be performed in neutral and abducted positions. Etiologies of compression may be identified, particularly osseous abnormalities such as cervical rib or post-traumatic osseous remodeling. CT angiography (CTA) may be performed to delineate the subclavian artery and may preclude the need to perform more invasive catheter directed diagnosis. Similar to CT and CTA, MRI and MR angiography (MRA) may be

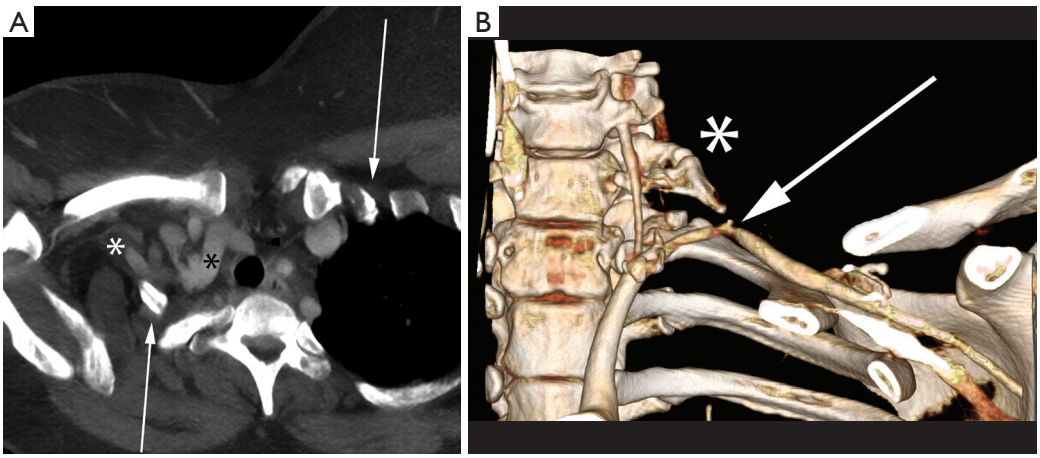

Figure 2 A 41-year-old female presented with lifelong weakness with pain and paresthesia in her right arm when elevated which did not respond to cervical discectomy or carpal tunnel release surgery. Examination of prior CT imaging revealed bilateral cervical ribs (image provided in *Figure 1*), raising suspicion for thoracic outlet syndrome. A thoracic outlet protocol CT angiogram demonstrate was performed. (A) 20 degrees coronally-obliqued axial maximum intensity projection demonstrates a dorsally tortuous course of the proximal right subclavian artery (black *) due scalene attachments to the free floating right cervical rib (arrow, patient right) resulting in superior displacement and narrowing of the scalene triangle. Note the severe stenosis of the right subclavian artery within the scalene triangle (hypoenhancing segment between the two asterisks) with distal reconstitution (white *). The fibrocartilaginous attachment of the articulating left cervical rib to the sternum is also noted (arrow, patient left), and similar subclavian arterial narrowing was observed on the left in this patient, although less severe (not imaged). (B) 3D volume rendering illustrates the narrowing of the left subclavian artery (arrow) and cervical rib (*). A diagnosis of arterial thoracic outlet syndrome was made, which did not respond to botox injections to the scalene and pectoralis minor musculature. Patient reported significant improvement of her symptoms following an uneventful resection of the right cervical rib.

used to evaluate the thoracic outlet. MRI is particularly useful in identifying soft tissue etiologies such as muscular hypertrophy or fibrous scarring. Images may be obtained first in the abducted position. If there is no evidence of narrowing, then further imaging in the neural position can be deferred. Catheter-based arteriography is the gold standard for assessing aTOS. Contrast material is injected from the aortic arch to assess for extrinsic compression of the subclavian artery in the neutral and abducted positions of the upper extremity. Other findings on angiography include subclavian artery aneurysm, thrombus, occlusion, dissection and distal emboli to the digits. Patients typically undergo a combination of these imaging modalities in diagnosis and planning for management of aTOS.

Management

Management of aTOS is not standardized and is largely dependent on the etiology. Options include conservative management or surgery which include rib resection, surgical bypass, scalene resection, as well as endovascular repair (4,7,9).

Conservative therapy

Conservative therapy for aTOS is not well studied due to scarcity of the disease coupled with diagnostic nuances, however, the available data suggest approximately half of patients with TOS benefit from at least 6 months of physical therapy with a small subset—around 5%—who actually worsen from physical therapy (10). Thus, the initial intervention for aTOS is frequently thrombolysis/thrombectomy either endovascular or surgical followed by maintenance anticoagulation (7).

Surgical intervention

Surgical intervention aims to decompress the neurovascular bundle and reconstruct involved vessels. This usually involves rib resection with complete anterior and middle scalenectomy, the hallmarks of surgical thoracic outlet decompression for bony abnormalities. Frequently the artery distal to the stenosis is abnormal as well, and almost 70% of patients undergo direct subclavian artery reconstruction with resection and interposition bypass graft

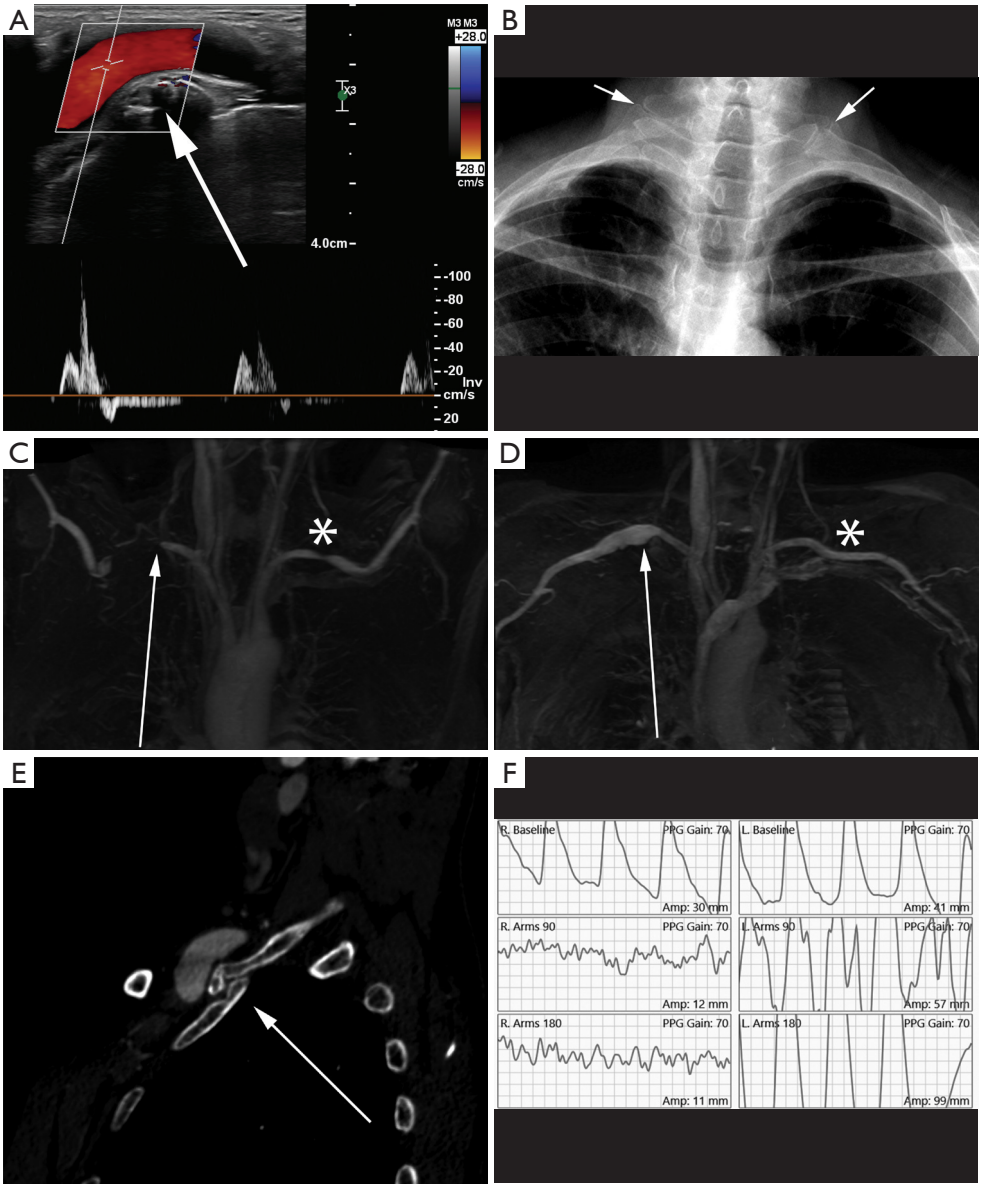

Figure 3 A 57-year-old woman presented with a pulsatile mass in the right supraclavicular region. (A) Ultrasound examination of the area of concern demonstrates a relatively superficial course of the right subclavian artery as it courses over what appears to be a bony rib (arrow). (B) Chest radiograph confirms the presence of bilateral cervical ribs in this patient (arrows). 3D maximum intensity projection (MIP) image from a gadolinium enhanced MR angiogram demonstrates occlusion of the right subclavian artery (arrow) in the abducted position (C). Note is also made of moderate narrowing of the left subclavian artery in the abducted position (*). Following repositioning of the arms to the adducted position (D), there is reconstitution of flow within the right subclavian artery (arrow), which demonstrates marked post-stenotic dilatation (arrow). The narrowing of the left subclavian artery was likewise relieved (*). (E) CT angiography further demonstrates the post-stenotic dilatation as the subclavian artery passes anterior to a pseudarthrosis (arrow) of the right cervical rib and 1st thoracic rib. (F) Pulse volume recordings demonstrate obliteration photoplethysmographic waveforms in the right arm in the 90 and 180 degree positions. These findings are consistent with arterial thoracic outlet syndrome.

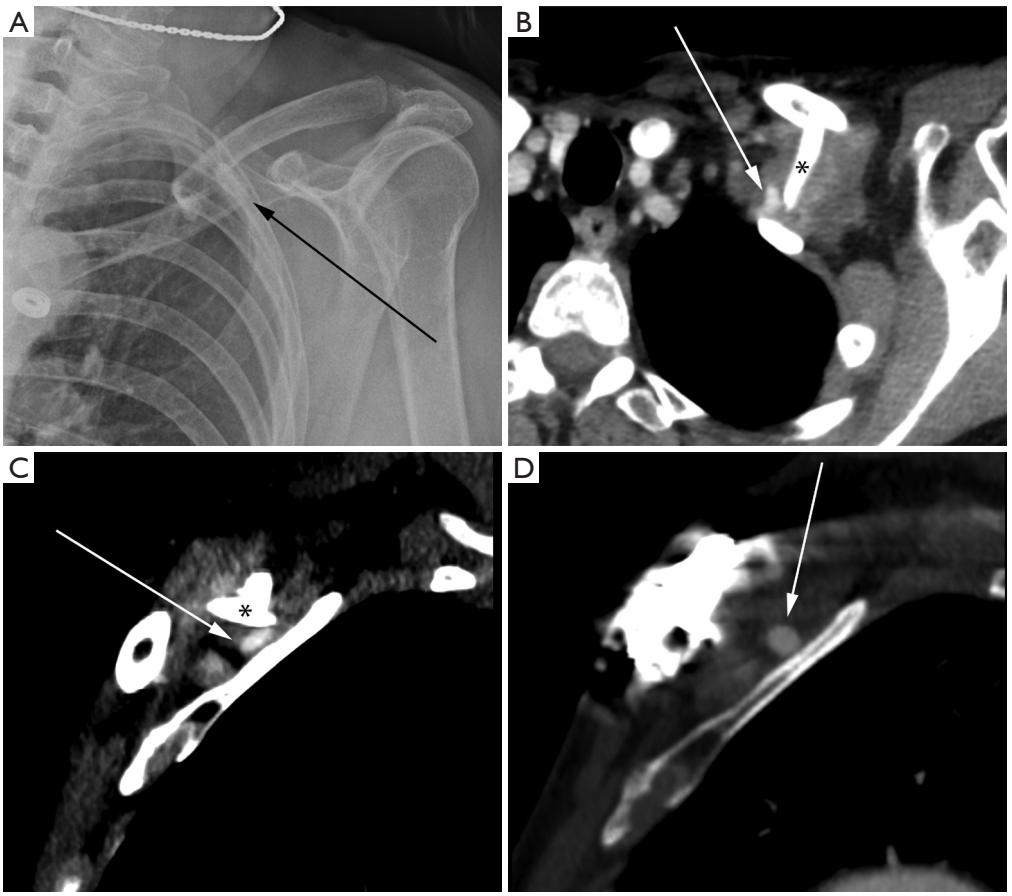

Figure 4 A 67-year-old female with concern for neurovascular injury after a fall onto her left side. (A) AP radiograph demonstrates a displaced midshaft clavicle fracture (black arrow). (B) CT-angiogram demonstrates the comminuted midshaft clavicle fracture with proximal and distal shafts separated by a 3 cm anterior-posterior oriented intercalated fracture fragment (black *), which displaces the left subclavian artery posteriorly (white arrow). There is a surrounding hematoma imparting additional mass effect. (C) Oblique sagittal reconstruction through the thoracic outlet demonstrates deformation and narrowing of the left subclavian artery (white arrow) as it is compressed between the intercalated fracture fragment (black *) and subjacent first rib. Brachial plexus compression was likewise confirmed on concurrently performed MRI (not included). Patient subsequently underwent open reduction and internal fixation, and 3 month follow-up CT angiogram (D) demonstrates interval decompression of the subclavian artery (arrow).

placement either cryopreserved organic or woven Dacron. Surgical endarterectomy and thromboembolectomy are also occasionally utilized during reconstruction. Post-operative outcomes demonstrate improvement of nearly 50% in terms of function and pain at 4 years (7).

Cervical rib resection

The incidence of cervical ribs in the general population is between 0.2–1% and classically defined as ribs arising from C7, occasionally crossing the thoracic outlet and fusing with the first rib resulting in vascular TOS (11). Nearly half of patients presenting with aTOS have cervical ribs. Surgical options include first rib with cervical rib resection either via transaxillary or supraclavicular approaches with possible aneurysm resection and bypass graft placement (12). There are no distinct advantages or disadvantages to either approach although there are studies suggesting that supraclavicular approaches are associated with longer operative times (13).

First rib resection and scalene resection

Anterior scalene muscle hypertrophy is linked with aTOS due to chronic compression of the subclavian artery as it traverses

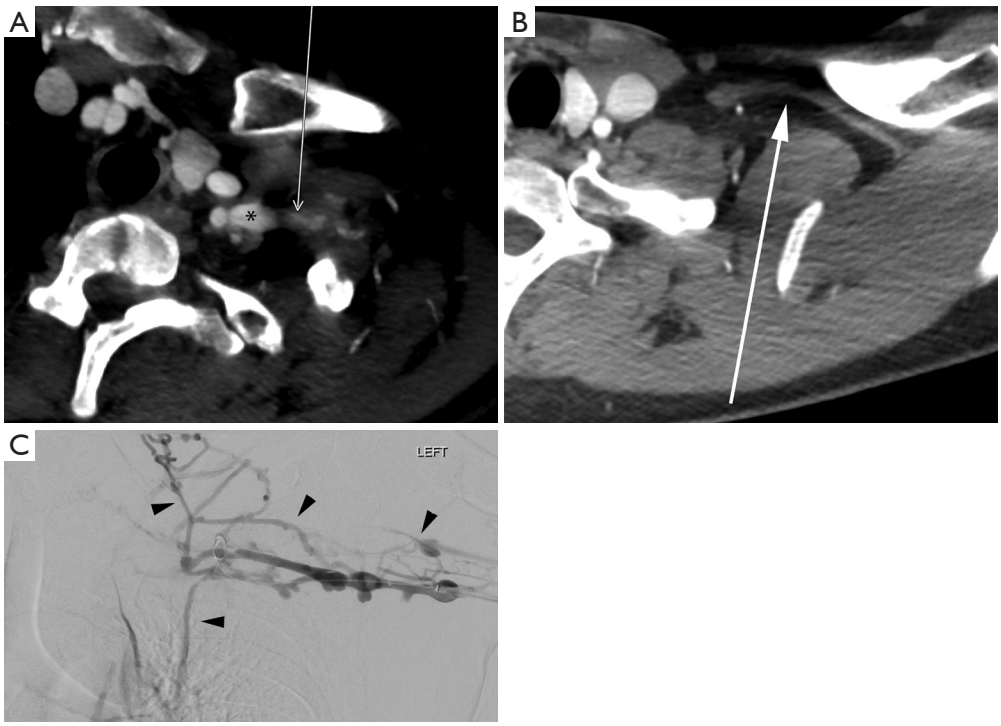

Figure 5 A 25-year-old patient with pain and paresthesia involving the left arm. (A) Oblique axial image in the neutral position demonstrates moderate to severe narrowing of the left subclavian artery within the scalene triangle (arrow). Note the difference in caliber and opacification relative to the proximal segment of the left subclavian artery (*). Findings confirmed a diagnosis of arterial thoracic outlet syndrome. Supernumerary ribs were not present. (B) CT images of the left subclavian vein from the same study likewise demonstrates significant attenuation in caliber. (C) Follow-up digital subtraction venogram demonstrated non-visualization of the left subclavian vein in neutral and stress positions with extensive shoulder collaterals (arrowheads), confirming a diagnosis of concomitant venous thoracic outlet syndrome. Patient was successfully treated with first thoracic rib resection.

the scalene triangle. Although there are no specific size criteria, there is a link between increased type 1 fibers (those used for tonic contracting) and patients with aTOS (14). Anterior scalenectomy is routinely performed with first rib resection for vascular TOS. Following exposure of the thoracic outlet, the anterior scalene muscle is transected and the first rib removed. Recovery takes approximately 8–12 weeks and the most common complication is pneumothorax (15).

Endovascular reconstruction

The current mainstay of aTOS therapy is open surgical repair however, concurrent endovascular reconstruction of the subclavian artery can be used to prevent downstream sequelae of aTOS which include functional deficiency and amputation from distal arterial embolization. Reconstruction is indicated in patients with subclavian aneurysms and endovascular stent graft reconstruction can be effective although current literature is limited (16).

Conclusions

The diagnosis and management of arterial thoracic outlet syndrome remains challenging. Patients may present with acute or chronic symptoms of arterial compromise, however in many patients the constellation of symptoms may reflect concurrent nTOS and vTOS in addition to aTOS. Diagnosis is made from physical exam and imaging findings with catheter angiography giving definitive evaluation of arterial compression, occlusion, aneurysm formation or dissection. Initial management is conservative, however in patients with continuing or progressing symptoms, surgical decompression may be used to treat aTOS. In patients who present with acute thrombosis, emergent catheter directed thrombolysis may be necessary to treat limb threatening ischemia. Catheter directed therapy may be performed as

an adjunct to, or after, open surgery to identify and treat subclavian arterial pathology. aTOS can be a debilitating disease however appropriate intervention can lead to improved functional and pain scores.

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Footnote

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