BRIGHAM AND WOMEN'S HOSPITAL
A Teaching Affiliate of Harvard Medical School
75 Francis St., Boston, Massachusetts 02115

Department of Rehabilitation Services
Physical Therapy

Standard of Care: Vascular Thoracic Outlet Syndrome (non-operative)

Case Type / Diagnosis:

Thoracic outlet syndrome (TOS) is described by a cluster of symptoms in the upper extremity. These include: pain in the shoulder and proximal upper extremity with or without neck pain, paresthesias and/or numbness into the distal upper extremity and hand, fatigability, swelling, discoloration, and Raynaud's phenomenon. There have been four symptom patterns described: upper plexus, lower plexus, vascular, and mixed. [33] Lower plexus symptom patterns are the most common. [33]

Generally, TOS is the result of compression of the neural and/or vascular structures between the interscalene triangle and the inferior border of the axialla. The structures that can cause compression of either neurological and/or vascular structures include: pectoralis minor tendon hypertrophy, clavicular deformity, cervical ribs, anomalous fibromuscular bands, or hypertrophy/injury to the scalene musculature. Developmental anatomical anomalies have been shown to be present in individuals that have symptoms of TOS. Makhoul and Machleder [25] reported in 1992 that 66% (132 of 200) of patients who underwent transaxillary surgical procedures for rib resection for the treatment of TOS had anatomic anomalies. They reported 17 cases of cervical or first thoracic rib abnormalities and 20 supernumerary scalene muscles. Incidentally, they also found 86 individuals with scalene and 39 with subclavius muscular developmental variations with regards to their insertions.

The terminology TOS was first introduced by Peet in 1956.[36] In 1958, Rob [39] first described TOS symptoms as arising from either compression of the brachial plexus and/or the subclavian vessels in the thoracic outlet region.
However, the first reports of cervical rib compression dates back to the Second Century AD. [1] In 1814, Coote was unsuccessful in resecting the cervical rib in a case of TOS. [9] It was not until 1905 that the first successful cervical rib resection was performed on a patient with TOS and a subclavian artery aneurysm. [31] In 1920, Law first described the possibility of soft tissue structures causing TOS. [19] The role of the scalene muscle in TOS was investigated in 1927. [1] The first successful surgical resection of the anterior scalene muscle was performed by Oschner et al [34] in 1935.

The diagnosis of TOS depends heavily on the subjective rather than objective criteria. Commonly the distinction between vascular thoracic outlet syndrome (VTOS) and neurogenic thoracic outlet (NTOS) is made. Hence published results of both conservative and surgical management vary greatly. [8] Ancillary studies are most helpful to rule out other conditions rather

than confirm the diagnosis of TOS. The diagnosis of VTOS is typically suspected by history and clinical presentation, and is confirmed by angiography or venography.

Vascular TOS:

There can be VTOS with venous thrombosis, and there can be venous thrombosis without VTOS. Venous thrombosis of the axillary and subclavian veins occurs in 2 per 100,000 individuals and accounts for less than 2% of all deep venous thrombi. [12, 21, 41] Infrequent immobilization, the presence of fewer valves, and decreased hydrostatic forces are common reasons for such a low incidence rate of thrombosis in these veins.[17, 37]Axillary-subclavian venous thrombosis (ASVT) is typically referred to as either primary or secondary. [35, 37] Primary thrombosis commonly occurs in the young, active, and healthy individual and can be the result of effort thrombosis [15] as the product of repetitive and/or strenuous activity. [40] Secondary thrombosis occurs in those with recognized thrombosis risk factors, usually in the older, more sedentary, and sicker individual.

Primary thrombosis is commonly found in individuals that have underlying anatomical abnormalities, which leads to venous compression at the site of the first rib and clavicle.[29, 43] Individuals with severe ASVT will present with VTOS symptoms of swelling of the upper extremity, with distended, prominent veins in the hand and forearm and possibly prominent veins along the chest. Other signs include bluish discoloration and a tender prominent cord in the arm, axilla, or neck. Occasional neurological signs (tingling or numbness) of the arm and hand may be due to the pressure of edema (accumulation of fluid) rather than nerve damage.

The presence of TOS symptoms may or may not be the result of vascular compromise. VTOS is less common than NTOS, but may result in significant long-term disability. [16]Individuals affected by VTOS are generally young, active, and have very few co-morbidities. [42] VTOS can exhibit either signs of arterial impingement/compression or venous obstruction. The latter is more common. It has been reported that the most common causes of arterial compression are from either a long cervical rib or an anomalous first rib. [10] Claudication, vasomotor phenomena, digital gangrene, limb heaviness, and acute limb-threatening ischemia are common presenting symptoms of arterial compression.

Potential Imaging Studies: These studies are commonly used (either individually or in combination) by physicians for the work-up of VTOS.

Radiographs:

- Cervical: May demonstrate a skeletal abnormality.
- Chest: May demonstrate a cervical or first rib (elevated or enlarged), clavicle deformity, and pulmonary disease.

Angiography/ Venography: Establishes the diagnosis of axillary-subclavian deep venous thrombosis. Angiography/venography is an x-ray method in which contrast material is injected into a blood vessel to visualize it. The physician may perform a complete examination by injecting contrast material into the affected arm and also rotating the arm to provoke compression of the vein. Indications for angiography/venography include evidence of peripheral emboli in the upper extremity and suspected subclavian stenosis or aneurysm. Conventional angiography is typically performed when surgical intervention is considered in order to confirm the extrinsic compression of the artery. MR angiography is a non-invasive approach and allows for a good evaluation of the subclavian artery in both adducted and abducted positions of the arm.

Color flow duplex scanning (ultrasound): Color-flow duplex ultrasonography (CDS) assesses the presence and severity of stenosis and yields a combination of anatomic and hemodynamic information. CDS allows veins to be surveyed longitudinally and facilitates the identification of veins. It also decreases the need to assess Doppler flow patterns and venous compressibility. [28]

Magnetic Resonance Imaging(MRI): MRI is commonly used for vascular imaging. Many different MRI techniques are used and each exploits different properties of blood flow to achieve contrast. Phase display imaging has proven useful in differentiating signal of slow flow from that of intravascular thrombus. Imaging of peripheral vessels can be achieved with gradient refocused sequences, which provide bright intravascular signal over a wide range of flow velocities. These sequences may be combined with subtraction strategies to eliminate the signal from stationary tissues in order to generate an angiographic image. The advent of three-dimensional MR angiographic imaging techniques provides an effective way to display peripheral vessels [20]

Methemoglobin is the product of a stage of a blood clot that reflects the oxygenation state of hemoglobin within the red cells. Methemoglobin acts as an endogenous contrast agent. Using a T1-weighted magnetic resonance sequence (Magnetic Resonance Direct Thrombus Imaging, MRDTI) methemoglobin is identified as a high signal. Subactute thrombosis can be identified by MRDTI. [30]

Demondion et. al [13] has demonstrated that MRI is helpful in identifying the location and cause of arterial or nervous compressions. In patients with TOS:

- Thicker subclavius muscle in both arm positions ($P < .001$).
- Wider retropectoralis minor space after the postural maneuver ($P < .001$).
- Venous compressions after the postural maneuver was observed in 63% of patients at the prescalene space.
- Venous compressions after the postural maneuver was observed in 61% of patients at the costoclavicular space.
- Venous compressions after the postural maneuver was observed in 30% of patients at the retropectoralis minor space.
- Arterial compressions were found in 72% of patients.
- Nervous compressions were seen in only 7% of patients.
- With the exception of venous thrombosis, vasculonervous compressions were demonstrated only with arm elevation.

Treatment:

Typically, the initial management for individuals with TOS is nonoperative with an emphasis on rehabilitative exercises. Surgical indications include: acute vascular insufficiency, progressive neurologic dysfunction, and/or unmanageable pain that failed conservative treatment. There are many surgical techniques; typically they involve the release and/or removal of the structures that cause compression (i.e. scalene/pectoralis minor muscle release, first rib resection, cervical rib excision, and resection of fibromuscular bands).

In terms of surgical management of TOS, careful selection of patients is required for satisfactory surgical results. A well-coordinated team of thoracic surgeons, neurologists, and physical therapists is key. [3] In addition, in more severe cases it has been shown that physical therapy cannot replace surgery. [2] Optimal treatment of TOS, either neurogenic or vascular, is highly controversial, especially regarding the role of surgery.

Surgical decompression for patients with TOS has been shown to be both an efficient and dependable treatment intervention; however, results worsen over time. [4] It has been reported that there is no significant difference in terms of relief of symptoms in postoperative outcomes between individuals who have had excision of either a cervical rib or of a first rib. [11] Landry et al. [18] reported on their series of 79 patients at mean follow-up of 4.2 years. Of the 79 patients, 15 had a first rib resection and 64 were managed conservatively. They found that most patients, who were conservatively managed, in their nonrandomized series, returned to work and had significant improvement in symptoms. In contrast, those individuals who underwent a first rib resection did not have an improved functional outcome.

This Standard of Care outlines the conservative physical therapy evaluation and management of a patient with VTOS.

Possible ICD.9 codes:

353.3 Thoracic Root Lesion
286.5 Hemorrhagic disorder secondary to circulation
453.8 Embolism and Thrombosis, Unspecified
671.4 Deep Phlebothrombosis

Indications for Treatment:

1. Upper Extremity edema
2. Impaired Range of Motion: Shoulder / Upper Extremity
3. Impaired Function: Shoulder / Upper Extremity
4. Poor Posture
5. Weak posterior (scapular, shoulder, trunk extensors) musculature
6. Tight anterior shoulder and chest musculature
7. Pain

Precautions for Treatment:

- No contraindications for physical therapy interventions, if the patient has been seen by BWH/F Cardiology/Vascular Surgery/Internal Medicine and have already been diagnosed with Vascular TOS and have begun on medical management. Attention should be directed to any possible worsening of vascular symptoms of the upper extremity as a result of increased upper extremity use (i.e. diminished pulse, discoloration, pain, etc). In addition, awareness of major warning signs of a recurrent DVT or PE should be continuously screened for: increased swelling of the upper extremity, increased redness of the upper extremity, return of other original symptoms (pain, heaviness of upper extremity), and shortness of breath.

- If suspected diagnosis of Vascular TOS upon physical therapy examination of the upper quarter, then immediately collaborate with the referring physician, nurse practitioner, physician's assistant, etc. for proper diagnosis and timely intervention. After collaboration with referring source(s), appropriate referral(s) will be made.

Examination:

Medical History: Review the Rehabilitation Department's medical history questionnaire (on an ambulatory eval), patient's medical record (during the inpatient stay) and medical history reported in the Hospital's Computerized Medical Record. Review any diagnostic imaging, tests, work up and operative report listed under LMR. Thoroughly review the attending physician's note(s) to determine underlying involved structure (which vascular tissue(s) are compromised).

History of Present Illness: Interview the patient to review history and any relevant information. If the patient is unable to give a full history, then interview the patient's legal guardian or custodian. Determine if any past injuries have taken place. Typically patients with VTOS have no previous injury, and symptoms are insidious in nature.

Social History: Review the patient's home, work, recreational, and social situation. Areas to focus on are upper extremity weight-bearing activities, excessive reaching, lifting, or carrying loads with upper extremities.

Medications: The physician may have prescribed anti-coagulation medication. This might be enoxaparin, dalteparin, fondaparinux, or warfarin, with or without concomitant aspirin or other nonsteroidal anti-inflammatory medication. If warfarin is prescribed, determine from the medical record or from the patient's health care provider what the target INR has been and the duration of warfarin administration. Patient's INR levels should be monitored via the lab results section in LMR to ensure adequate anti-coagulation.

Examination (Physical / Cognitive / applicable tests and measures / other)
This section is intended to capture the most commonly used assessment tools for this case type/diagnosis. It is not intended to be either inclusive or exclusive of assessment tools.

Pain: As measured on the Visual Analog Scale/Verbal Rating Scale/Numerical Rating Scale, activities that increase symptoms, decrease symptoms, location of symptoms and irritability level.

Visual Inspection: Attention to the healing of the incision (if they have any interventional treatment for the management of the venous thrombosis), ensuring there are no signs of infection. In addition, visual inspection of the involved upper extremity in regards to edema, discoloration, and overall appearance.

Palpation: Comparison of involved and uninvolved extremity.

- Palpate entire shoulder girdle and upper extremity. Focus on presence and extent of musculature atrophy and swelling.
- Pulses: Carotid, Brachial, and Radial.
- There may be pain with palpation of the scalene musculature, the sublavius muscle as it attached to the 1st rib, and also along the brachial plexus.
- There maybe a positive Tinel sign over the superclavicular area at the insertion of the anterior scalene muscle.

Edema: Girth measurements to be taken on both involved and uninvolved upper extremity (in centimeters):

- Widest part of upper arm. (Document level as # of cm. distal to the tip of the acromion)
- Elbow (around the olecrenon process).
- Wrist
- Severe edema can be assessed using upper extremity volumetric measurements.

ROM:

- Initial ROM assessment of the cervical spine, involved shoulder, and entire upper extremity as compared to the uninvolved side.
- Muscle length testing: Pectoralis Major & Minor, Scalenes, Sternocleidomastoid.

Muscular Performance: Manual Muscle Testing (MMT) is used to get a baseline of a patient's strength. Particular attention should be placed on upper back strength and proximal shoulder strength. Likely poor strength in these areas will be present leading to poor posture and overall poor shoulder mechanics. Handgrip with dynamometer testing is recommended at baseline. Muscle weakness is typically not noted. However, if present, it is typically mild and most prominent in the thenar, hypothenar, and interosseous muscles inervated by the ulnar nerve.

Sensation: Hypestehesia may occure in the C8-T1 dermatomes. If sensation is found to be abnormal via objective dermatomal screen, further assessment would be indicated. Perform Tinel testing of ulna and median nerves; carpal tunnel compression. In addition, additional neural tension testing can be utilized. Documentation should be specific

regarding the point/ position of positive symptoms. Subsequent re-testing can be used to compare progress or decline of symptoms of irritability. A Semmes-Weinstein Monofilament Screen may be used in order to identify patients with peripheral nerve branch involvement as well as to track their progress. In general, monofilament testing has been shown to be a sensitive monitor of peripheral nerve function. [16, 24] The Semmes-Weinstein monofilaments have been shown to vary relatively little in terms of their application force. These forces are consistently reproducible over time in clinical testing.

Posture/alignment: Primary focus on sitting and standing upper quadrant and upper back posture. These patients tend to be at the extremes of rounded shoulders and forward head positions.

Breathing: With relaxed breathing the scalene musculature is active on inspiration through full inspiratory excursion. However, patients with TOS often are unable to keep the scalene musculature quiet during inspiration. Patient typically have difficulty with diaphragmatic breathing.

Neurodynamic Testing: The nervous system should be examined both functionally and specifically. Functional examination consists of having the patient elevate their arms with the elbow extended and with the elbow flexed. The point of tension is noted during the elbow range of motion. This is position is compared to specific examination of upper limb tension testing. There are 4 main tests that assess the extensibility of neural structures, with each one biasing a different aspect of the nervous system. Full description of each test can be found in Chapter 3: The Cervical Spine of Orthopedic Physical Therapy Assessment by Magee. [25]

Gait & Balance: Gross assessment to determine patient's safety and to ensure independence with transfers, gait, and stairs. Further in depth assessment to be conducted if impairments noted in screening.

Special Tests: Potential thoracic outlet special tests:

- Allen
- Wright
- Adson
- Halstead
- Roos
- Costoclavicular
- Hyperabduction

(Full description of each test can be found in Chapter 5: The Shoulder of Orthopedic Physical Therapy Assessment by Magee. [24])

A cluster of special tests is recommended when evaluating a patient with suspected TOS. Gillard et al. [14] prospectively assessed 48 patients with a clinical presentation of TOS. They used a number of standardized provocative tests (three of which were the

Adson, Hyperabduction Test, Wright), an electromyogram, a Doppler ultrasonogram, and a helical CT arterial and/or venous angiogram to evaluate for the presence of TOS. They found that the cluster of these provocative tests had mean sensitivity and specificity values of 72% and 53%, respectively, with improved values for the Adson test (positive predictive value [PPV], 85%) and the hyperabduction test (PPV, 92%). The more positive the provocative tests, the higher the specificity. Doppler ultrasonography visualized vascular abnormalities and supported the diagnosis in patients with at least five positive provocative tests. Electrophysiological studies were found to be helpful for differential diagnosis and for detecting concomitant abnormalities.

Differential Diagnosis:

- NTOS as a stand-alone issue or in conjunction with vascular compromise.
- Shoulder Pathology
- Pathologic Lesion (tumor/cyst/infection)
- Cervical Radiculopathy
- Brachial Plexus Neuritis / Injury
- Postural Palsy
- Raynaud Disease
- Ulnar Nerve Compression (at the elbow)

Referral Source Considerations:

- Patients referred from the BWH/F Cardiology/Vascular Surgery/Internal Medicine Service with a Diagnosis of Vascular TOS, have already been thoroughly worked up for Vascular TOS and begun medical management.
- Patient may have co-morbid issues that need to be considered.

Functional Assessment:

Use of a shoulder specific functional capacity questionnaire is recommended to establish early initial status and track progress.

Possible tools:

- Simple Shoulder Test (SST)
- American Shoulder and Elbow Surgeon's Shoulder Evaluation Short Form (ASES-SF)
- Shoulder Pain and Disability Index (SPADI)

The SST [22]and the ASES-SF[6], and the SPADI [38] are all standardized self- assessments of shoulder function and have been found to have fairly high responsiveness as well as high test-retest reliability as compared to other shoulder outcome tools. [5] The SST has a standardized response mean of 0.87, confidence interval 0.52, 1.22; while the ASES-SF had a standardized response mean of 0.93, confidence interval 0.57, 1.29. The intraclass correlation coefficients for the SST and ASES-SF are 0.99 and 0.96,

respectively. They both are very simple and quick for the subject and investigator to fill out. The SST has been shown to be sensitive for various shoulder conditions as well as sensitive to detect changes in shoulder function over time. [26, 27] The SPADI is another subjective questionnaire that has a pain and disability/function components. This scale uses a visual analog scale to measure pain while subjective questions are used to assess function of the shoulder. The pain and function components are weighted accordingly since there are 5 pain scales and 8 functional questions, and then the total score is computed by averaging the pain and functional score. With the SPADI, unlike the other outcome measures a higher value indicates greater pain and disability.

Evaluation / Assessment:

Establish underlying reason for need of Skilled Services.

Potential Problem List (Impairment(s) and/ or dysfunction(s))

1. Pain
2. Edema Involved Upper Extremity
3. Decreased ROM Shoulder / Upper Extremity
4. Decreased Strength Upper Back Musculature / Upper Extremity Musculature
5. Decreased Function as compared to baseline
6. Decreased Knowledge of Activity Modification
7. Decreased Knowledge of Rehabilitation Progression

Prognosis/Expected Outcomes: Literature Review:

Little is known about the overall long-term outcome of patients with Vascular TOS and conservative management. Please refer to previous literature review. Upon discharge from skilled physical therapy intervention, these patients continue on anti-coagulation management, under the direction of their health care provider. They are advised to continue with their home exercise program indefinitely.

The alternative to conservative (medical and physical therapy) management for patients with Vascular TOS is surgical intervention that resects the pathological symptom provoking structure (first rib, scalenes, or other muscular structures).

Goals of Intervention
Goals of intervention are individualized for each patient's current health status. Potential goal categories are:

1. Decrease Pain
2. Decrease Edema
3. Restore ROM
4. Restore Strength
5. Improve Posture
6. Improve Body Mechanics
7. Enhanced / Normalized Breathing
8. Increase Function
9. Independent with Home exercise program

Treatment Planning / Interventions

If cleared by the BWH/F Cardiology/Vascular Surgery/Internal Medicine Service, and currently being medically managed for VTOS, there are no restrictions in terms of aerobic and anaerobic exercise.

Established Pathway ___ Yes, see attached. _X_ No

Established Protocol ___ Yes, see attached. _X_ No

Interventions most commonly used for this case type/diagnosis.
This section is intended to capture the most commonly used interventions for this case type/diagnosis. It is not intended to be either inclusive or exclusive of appropriate interventions.

Typically these patients present with impairments that require:

- Patient / family education as below
- Potential use of modalities to assist with pain reduction
- A gradually progressed anterior (cervical spine and trunk) muscular stretching program
- Gradually progressed upper back strengthening program
- Joint mobilization as indicated (rib, glenohumeral, scapulothoracic, cervical spine joints)
- Restoration of shoulder / upper extremity ROM through Active / Passive ROM, Hold/Relax, Contract/Relax Techniques
- Establishment of appropriate diaphragmatic breathing
- Gradual functional activity progression including work and recreational activities.

Frequency & Duration

Initial physical therapy assessment should be completed as soon as possible (hopefully within 24 hours) of physician referral.

Outpatient Care: 1-2x week/ for 2-3 months as indicated by patient's status and progression.

Patient / family education

1. Instruction in HEP (home exercise program)
2. Instruction in correct posture
3. Instruction in appropriate breathing
4. Instruction in correct body mechanics
5. Instruction in pain control and ways to minimize inflammation
6. Instruction in activity level modification / joint protection
7. Distended or swollen veins may or may not resolve with treatment.

Recommendations and referrals to other providers.

None, except back to Attending Physician if issues arise, such as:

- Marked Increased swelling of the upper extremity
- Marked Increased redness of the upper extremity
- Return of any of the other original symptoms (pain, heaviness of upper extremity)
- Shortness of Breath

Re-evaluation / assessment

Standard Time Frame- 30 days or less if appropriate

Other Possible Triggers- A significant change in signs and symptoms

Discharge Planning

Commonly expected outcomes at discharge – Patient should be/have:

- Independent with Home Exercise Program
- Independent with self management of symptoms
- Independent with Posture correction
- Independent with correct Body Mechanics
- Full Shoulder / Upper Extremity ROM
- Upper back strength of all musculature of at least 4/5
- Shoulder / upper extremity musculature strength of at least 4/5
- Either completely resolved or minimized upper extremity edema
- Know the warning signs of a recurrent DVT or PE:
    1. Marked Increased swelling of the upper extremity

2. Marked redness of the upper extremity
3. Return of any of the other original symptoms (pain, heaviness of upper extremity)
4. Shortness of Breath

Transfer of Care – Possibly a physical therapist closer to where the patient lives, if traveling to BWH is too inconvenient for consistent rehabilitation care. In this case the therapist in the community will be given a copy of this standard of care to assist them in guiding the patient's treatment.

Patient's discharge instructions – Continue with individualized home program indefinitely to ensure maintenance of ROM, strength, posture and function.

Author:
Reg Wilcox III, PT
05/04

Reviewed By:
Kenneth Shannon, PT 6/04
Amy Bulter, PT 6/04
Heather Renick-Miller, PT 6/04
Sam Goldhaber, MD 6/04
Janice McInnes, PT 6/04

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