Current Management of Thoracic Outlet Syndrome

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Opinion statement

Thoracic outlet syndrome (TOS) is a condition caused by compression of the neurovascular structures leading to the arm passing through the thoracic outlet. There are three distinct types of TOS: neurogenic (95%), venous (4%–5%), and arterial (1%). Treatment algorithms depend on the type of TOS. Although statistically the most common type, neurogenic TOS can often be the most difficult to diagnose and treat. We have good follow-up data indicating that appropriately selected patients benefit from surgical intervention. Arterial and venous TOS often present more urgently with arterial or venous thrombosis. The thrombosis is typically recognized expeditiously by thorough history taking and physical examination, augmented by duplex ultrasonography. The restoration of blood flow, be it venous or arterial, often can be accomplished readily by thrombolysis. The key, however, comes in diagnosing the underlying structural component involved in the development of symptoms. To prevent recurrence, patients must undergo first rib resection and anterior scalenectomy, as well as resection of any rudimentary or cervical ribs. In the case of arterial TOS, the subclavian artery often requires reconstruction as well. Regardless of the type of TOS encountered, proper treatment requires a multidisciplinary approach.

Introduction

Historically, thoracic outlet syndrome (TOS) has been a controversial syndrome in terms of its causes, diagnosis, and management. Around 150 AD, Galen first described the cervical rib in human dissections [1]. As his work had fallen out of favor during the Dark Ages, Vesalius, who revived much of Galen's work, again described the cervical rib among human cadavers [1]. In 1821, Sir Astley Cooper [2] observed that a subclavian artery occlusion was caused by compression due to a cervical rib, which thus came to be known as cervical rib syndrome. By 1927, the first rib was implicated in neurogenic TOS [3]. In 1935, Ochsner et al. [4] described the *scalenus anticus syndrome*, and an association was first made with trauma. In 1943, the term *costoclavicular syndrome* was used to describe compression of the subclavian artery and vein [5]. The term *thoracic outlet syndrome* was first introduced in 1956 by Peet et al. [6], who also described a therapeutic exercise regimen in the management of the syndrome.

As the differing views of the anatomic causes of TOS changed over the years, so did the surgical approaches to its management. In 1861, Coote [7] resected the first cervical rib for relief of arterial compression. Murphy [8] performed a first rib resection in 1910 for neurogenic TOS. Adson and Coffey [9] performed the first scalenectomy in 1927, without rib resection. Later it was realized that the division of the muscle alone led to recurrent symptoms. The posterior approach to first rib resection was described by Clagett [10] in 1962, and the transaxillary approach to first rib resection

Table 1. Thoracic outlet syndrome: types and characteristics

Characteristic	Neurogenic TOS (most common type)	Venous TOS (subclavian vein thrombosis)	Arterial TOS (subclavian artery thrombosis)
Patient's sex	Female (3.5:1)	Male	Female/male equally
Patient's age, y	20–40	20–30	20–30
Risk factors	Repetitive movements Previous trauma	Strenuous work Athletics	Vigorous arm activity
Symptoms	Pain down arm, forearm, ring finger, and little finger Paresthesias (pain, numbness) at night Arm/hand weakness Arm/hand swelling Loss of dexterity Cold intolerance Headache	Pain in affected arm, often associated with strenuous work Arm/hand swelling Veins of shoulder and chest appear more visible Hand/arm appears blue	Pain Claudication Hand appears white Hand/arm cool Decreased pulse
Laboratory studies	None	Hypercoagulable studies	Hypercoagulable studies
Imaging studies	Chest radiography	Chest radiography Duplex/ultrasound Venography	Chest radiography Duplex/ultrasound Arteriography
Other studies	Nerve conduction study	Nerve conduction study	Nerve conduction study
Consultations	Vascular surgeon Physical therapist Pain management specialist	Vascular surgeon Hematologist	Vascular surgeon Hematologist
Treatment	PT Lidocaine blocks Botox* blocks Surgical intervention† Postoperative PT† Medication	Anticoagulation Surgical intervention Venography/venoplasty Postoperative PT	Anticoagulation Surgical intervention Arteriography/arterioplasty Postoperative PT
Prognosis	Good	Good	Good
Recovery time	Longer period of time (Time needed depends on duration of symptoms before treatment and commitment to PT)	Quick	Quick

*Brand of botulinum toxin type A manufactured by Allergan, Irvine, CA.
†Not all neurogenic TOS patients need or are candidates for surgery.
PT—physical therapy; TOS—thoracic outlet syndrome.

was described by Roos [11] in 1966. Other surgical approaches include infraclavicular as well as combined techniques using transaxillary first rib resection and transcervical scalenectomy [12,13].

TOS is divided into three distinct subtypes: neurogenic, venous, and arterial. Symptoms are produced relative to which of the neurovascular structures is compressed in the thoracic outlet (Table 1). In this regard, the scalene triangle is the most important anatomic space in the thoracic outlet. The anterior and middle scalene muscles comprise the sides of the scalene triangle, with the first rib forming the base of the triangle. The anterior and middle scalenes both arise from the lower cervical spine. Through this relatively small space pass the subclavian artery and vein and the five nerve roots of the brachial plexus. These muscles can potentially scar and/or hypertrophy with repetitive motion or trauma, further contributing to compression of the neurovascular structures. The subclavius muscle, which can be found between the clavicle and the first rib, also may compress the subclavian vein, contributing to venous TOS. This muscle often is hypertrophied in athletes presenting with effort thrombosis [14••].

A combination of congenital anatomic predispositions and acquired extrinsic factors results in the spectrum of symptoms associated with TOS. Cervical ribs, which are congenital, can predispose to all three types of TOS. They have an incidence of approximately 1% in the general population, and 50% are bilateral [15,16]. Although most cervical ribs are asymptomatic, they occur in up to 11% of patients undergoing surgical decompression for TOS [17,18]. A long C7 transverse process or bifid first rib also may predispose to TOS, but these are even less common. Particularly in neurogenic TOS,

congenital fibromuscular bands often are observed—in more than 80% of patients, according to some authors [19]. These congenital bands and ligaments have been categorized into nine different types by Roos [20]. Anatomic variations in the size and insertion points of the scalene muscles also have been implicated.

In addition to these (and other) predisposing anatomic factors, trauma and repetitive motion also play a role in the development of TOS. Some authors have reported that up to 80% of TOS patients have a history of trauma to the neck or shoulder area [21]. Repetitive motion due to athletic activity (eg, swimming or weightlifting) or occupational environment (eg, poor posture and/or prolonged computer use) also may contribute to TOS.

NEUROGENIC TOS

Symptoms attributable to neurogenic TOS are caused by compression of the roots of the brachial plexus and are much the same as those experienced with nerve compression in other parts of the body. Pain, paresthesias, and weakness in the pattern of the affected nerve roots are common. Pain and/or point tenderness in the anterior or posterior shoulder region, as well as the neck and occipital or mastoid region, is also a common finding. Headaches also may be present. Surgical intervention for neurogenic TOS is reserved for patients who have failed physical therapy, which typically lasts for 8 weeks. Physical therapy is aimed at improving posture and increasing range of motion. Because these patients often have repetitive motion as a consequence of their vocation, therapy is also directed toward a more ergonomic work environment. If physical therapy is not successful, a CT-guided anterior scalene block is performed with local anesthetic. The block causes relaxation of the anterior scalene muscle, allowing the first rib to drop, which should relieve tension on the brachial plexus. Symptomatic relief as the result of a scalene block is a reliable indication that first rib resection and anterior scalenectomy will be of benefit. We also have found that CT-guided botulinum toxin type A (Botox; Allergan, Irvine, CA) injections provide symptomatic relief for at least 1 month, and often up to 3 months [22]. These treatments may be used as a bridge to surgery or as an adjunct to nonoperative treatment with physical therapy. We do not offer more than two injections because the muscle and surrounding tissue tend to scar and the long-term effects of repeated Botox injections are not known. When neurogenic patients are appropriately selected for surgical intervention, we have shown good outcomes in terms of quality-of-life improvement [23••].

VENOUS TOS

The most common presentation of venous TOS is acute effort thrombosis of the axillosubclavian vein, also known as Paget-Schroetter syndrome. Patients with this syndrome, who typically are athletes, may present emergently with an acutely swollen, often discolored upper extremity. Duplex ultrasonography is the diagnostic test of choice and will reveal partial or complete thrombosis of the axillary and/or subclavian veins. Therapeutic anticoagulation followed by venography and catheter-directed thrombolysis is appropriate, provided it is instituted within 2 weeks of the onset of symptoms. Venous angioplasty or stenting should not be attempted in this setting because the mechanism of stenosis is extrinsic compression from the first rib. After successful thrombolysis, therapeutic anticoagulation is maintained on an outpatient basis. Days to weeks later, the patient will undergo first rib resection and anterior scalenectomy. To avoid bleeding complications, no anticoagulation is used until 3 days after surgery. Therapeutic low molecular weight heparin injections are given until 2 weeks later, at which time the patient will undergo repeat venography. At this time, stenotic areas can be dilated effectively as the extrinsic compression has been relieved. If the vein has remained patent and no intervention is required, then anticoagulation is discontinued. If the vein has rethrombosed, repeat thrombolysis may be performed and anticoagulation continued with warfarin, with an international normalized ratio goal of 2 to 3. The patient is followed up long term with duplex ultrasound monthly, and anticoagulation is continued as long as areas of increased velocities in the subclavian vein are visualized during arm abduction, up to 6 months. In our series of more than 80 patients with venous thrombosis, we observed that after 6 months, all remained patent; therefore, we do not anticoagulate beyond this time. The final important consideration is a thorough hypercoagulable workup, which is described later.

ARTERIAL TOS

The least common type of TOS, accounting for less than 1% of cases, arterial TOS is often associated with a cervical rib. Patients present with varying degrees of ischemia in the arm or hand. Ischemia may be caused by external compression or from emboli arising from an associated subclavian artery aneurysm. Ischemia may manifest with many signs and symptoms, including weakness, paresthesias, pain, pallor, and diminished pulses distally, especially with activity or positional changes. A positive Adson test helps establish the diagnosis but is not pathognomonic. Chest radiography is recommended to detect a cervical rib, which frequently is present in cases of arterial TOS. Duplex ultrasound also is essential to assess the presence and degree of arterial stenosis and to detect subclavian artery aneurysm [24]. In the case of arterial TOS, in addition to first rib resection and anterior scalenectomy, the artery frequently must be replaced with saphenous vein or prosthetic material to maintain perfusion to the upper extremity.

Treatment

Lifestyle

- Modification of daily activities, including minimizing repetitive motion, overhead work, and weightlifting, may improve symptoms.
- Proper posture when walking, sitting, and standing promotes relaxation of the neck muscles and may help decompress the thoracic outlet.
- Exercise directed toward improved range of motion and flexibility also may provide symptomatic improvement.

Pharmacologic treatment

- Nonsteroidal anti-inflammatory drugs may provide symptomatic relief but should be used with caution in patients with a history of peptic ulcer disease or bleeding problems.
- The use of opioid analgesics should be discouraged in general because of the often chronic nature of pain associated with TOS and the potential for addiction.
- Muscle relaxants may provide relief in acute exacerbation, but classes such as benzodiazepines must be used with caution because of their addictive potential. Muscle relaxants with lower addictive potential, such as carisoprodol and metaxalone, can provide symptomatic relief and may be used to augment physical therapy.
- Tricyclic antidepressants and selective serotonin reuptake inhibitors can provide relief from neuropathic pain.

Interventional procedures

Arteriography

	Arteriography is indicated only for patients in whom arterial involvement is suspected.
Standard procedure	A transfemoral approach typically is used so that arteriography can be performed with the arm in a variety of positions (eg, adduction, abduction, above the head).
Contraindications	Few, but contrast allergy and renal insufficiency must be taken into consideration. Also, patients who are anticoagulated should have their anticoagulation stopped soon enough to allow for normal coagulation profiles.
Complications	Contrast nephropathy, contrast reaction, access complications (ie, bleeding, thrombosis, or embolism).

Venography

	Venography is indicated only for patients in whom venous involvement is suspected. It may be combined with thrombolysis in acute axillosubclavian thrombosis or Paget-Schroetter syndrome.
Standard procedure	Typically performed from a brachial approach, using ultrasound-guided venous access.
Contraindications	Similar to those for arteriography. If thrombolysis is considered, then absolute contraindications include recent neurosurgery or head trauma, recent stroke, and active bleeding or recent gastrointestinal bleed. Major relative contraindications include recent cardiopulmonary resuscitation, recent major nonvascular surgery or trauma, uncontrolled hypertension, recent eye surgery, intracranial tumor, and puncture of a noncompressible vessel.

Complications	Similar to those for arteriography. Obviously, bleeding risk is much higher with thrombolysis; risks include hemorrhagic stroke in addition to hemorrhage from other sites.

CT-guided scalene muscle block

	CT-guided scalene muscle block has high sensitivity and specificity for diagnosing neurogenic TOS [25]. Also, patients experiencing symptomatic relief from scalene block with local anesthesia are more likely to have a good outcome with surgery.
Standard procedure	Used as a diagnostic tool if the provider is uncertain whether a patient has neurogenic TOS. Also, given its ability to reliably predict successful outcome with surgery, it can be used to convince a patient considering thoracic outlet decompression.
Contraindications	Allergy to local anesthetic agent.
Complications	Dysphagia (transient) and hematoma.

CT-guided scalene muscle Botox injection

	CT-guided scalene muscle Botox injection provides a longer period of relief compared with local anesthetic block of the scalene muscles. This approach can be used while patients are awaiting surgery or to augment conservative treatment with physical therapy.
Standard procedure	Good for patients who have had recent cervical spine or shoulder surgery, in whom thoracic outlet decompression should be delayed for at least a year.
Contraindications	Presence of infection at the planned injection site, history of allergic reaction or sensitivity to the injection, pregnancy (category C), breastfeeding, and history of neuromuscular disorders.
Complications	Dysphagia (may last 2–3 weeks) and hematoma.

Surgery

- The goal of surgery is to relieve compression of the neurovascular structures in the thoracic outlet. Arterial involvement often requires direct approaches for reconstruction in addition to decompression. Indications for surgery include vascular complications, failure of an adequate course of conservative therapy, and disability to the extent of interference with work or activities of daily living. There are multiple surgical approaches, each with specific advantages and disadvantages.

Transaxillary rib resection and anterior scalenectomy

	Transaxillary rib resection and anterior scalenectomy is probably the most common approach to thoracic outlet decompression and is our favored technique. This approach provides good visualization of the major structures through a cosmetically acceptable incision.
Advantages	Rapid and easy exposure of the first rib and anterior scalene with little manipulation of the neurovascular structures. Recovery is generally rapid, and the incision is cosmetically acceptable. Cervical ribs also can be removed with this approach. This is easily the best approach for venous TOS as it provides good visualization of the subclavius muscle, which tends to be hypertrophied in these patients.
Limitations	Lighting may be a problem in the relatively small, deep space of the axilla; however, this is overcome quite well with fiberoptic lighted retractors and use of a headlight. In cases of arterial involvement, adequate exposure of the subclavian artery for purposes of reconstruction is almost impossible.

Supraclavicular scalenectomy with or without first rib resection

	Supraclavicular scalenectomy with or without first rib resection provides good visualization of the thoracic outlet structures; however, because of the need to mobilize and retract these structures, the risk of injury is higher.
Advantages	Good visualization of the thoracic outlet structures and adequate exposure for arterial reconstruction, if necessary. Cervical ribs can be removed readily with this approach.
Limitations	Because dissection and retraction of the phrenic and long thoracic nerves and the brachial plexus are required, these structures are at higher risk of injury. The incision for this approach also tends to have less cosmetic appeal. If scalenectomy alone is performed without first rib resection, recurrence of symptoms has been shown to be higher.

Combined approach

	Another strategy combines the axillary approach for first rib resection and the supraclavicular approach for scalenectomy and arterial reconstruction. Although this combination approach is useful for patients with arterial involvement, it is not necessary for most TOS patients.
Advantages	Allows for thoracic outlet decompression and arterial reconstruction in one setting.
Limitations	Requires repositioning of the patient after the first portion of the procedure, prolongs operative time, and requires two incisions.

Posterior approach

	The posterior approach applies mainly to patients in whom the anterior or axillary approach is deemed more difficult or impossible, perhaps because of radiation-induced tissue fibrosis.
Advantages	May be beneficial for patients who have undergone a previous anterior or axillary approach for TOS and require reoperation, or for those who have had anterior chest wall radiation.
Limitations	Because it requires a posterior periscapular incision and division of the trapezius muscle for adequate elevation of the scapula, this approach is much more morbid in terms of pain and time to recovery. Visualization of the anterior structures is difficult, and arterial reconstruction is impossible.

Physical therapy

- Physical therapy is extremely important, particularly in the treatment of neurogenic TOS. It can be used as the sole treatment method, as some patients achieve relief of symptoms with exercise, strengthening, postural changes, and improved flexibility. If physical therapy alone fails and surgery becomes necessary, postoperative physical therapy is equally important for maintaining the long-term relief of symptoms.

Special points

A course of physical therapy consisting of three sessions per week for a total of 8 weeks is our standard before considering surgery for neurogenic TOS. Postoperatively, we prescribe the same frequency and duration of therapy to include increased range of motion, strengthening exercises, and soft tissue/scar tissue massage.

Cost/cost-effectiveness

$30 to $40 per session.

Pediatric considerations

- TOS is unusual in the pediatric population and is typically limited to patients with rudimentary or cervical ribs. Indications for surgery for children are typically the same as for adults, including vascular complications or significant disability that has not responded to conservative therapy.

Other considerations: hypercoagulable evaluation

- We have observed an increased frequency of hypercoagulable disorders in patients with Paget-Schroetter syndrome. Therefore, we now perform a hypercoagulable workup in all our venous TOS patients, as well as in arterial thrombosis patients. If the patient presents acutely, these laboratory values will be assessed initially. If the patient presents from another institution where he or she has already undergone thrombolysis and is being maintained on warfarin therapy, the hypercoagulable evaluation is done 6 months after discontinuation of warfarin. These laboratory tests include protein C and S levels, factor V Leiden mutation, prothrombin gene mutation 20210, antithrombin III level, antiphospholipid antibodies (lupus anticoagulant and anticardiolipin antibodies), homocysteine levels, and methylene tetrahydrofolate reductase (MTHFR) 677T variant. If the patient is confirmed to have a hypercoagulable disorder, then he or she is continued on lifelong anticoagulation.

Disclosures

No potential conflicts of interest relevant to this article were reported.

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have been highlighted as:
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