Thoracic Outlet Syndrome-An Old Challenge with a New Image

A Common Clinical Challenge

Physicians frequently face the common and challenging clinical problem of the patient with neck, shoulder, and arm pain. The problem lies in differentiating between cervical spine, musculoskeletal, and neurologic etiologies. The differential diagnosis is often confounded by superimposed psychosocial and emotional issues when the patient has been previously seen by multiple physicians and caregivers without resolution of their complaints.

Thoracic outlet syndrome (TOS) is one of the most confusing etiologies in this group of patients, and is much more common than previously recognized. In the past, the pathophysiology, diagnosis and true extent of TOS have generated considerable controversy. Today, there are many prestigious university medical centers and many esteemed researchers who have clarified this controversy, and who specialize in the diagnosis and treatment of patients with thoracic outlet syndrome.

This white paper explores the controversy of TOS, the anatomy and pathophysiology of TOS, the diagnostic challenges in patients with TOS, the treatment options in patients with TOS, and the use of new MRI technologies in the diagnosis of TOS.

Controversy

Many physicians are only briefly exposed to the concepts of thoracic outlet syndrome during medical school, or during their residency. Due to this brief exposure and to the complexity of thoracic outlet syndrome, many physicians recall the controversy more than they recall specific information about thoracic outlet syndrome. Fortunately, there is a very large body of medical literature that clarifies and confirms the existence, diagnosis, and treatment of thoracic outlet syndrome.
Thoracic outlet syndrome was first recognized more than 180 years ago. The history of thoracic outlet syndrome includes some of the most renowned names in medical history:

A. Sir James Paget is widely recognized as one of the founders of modern pathology, and was one of the most famous surgeons in London in the late 1800s. Sir Paget first described thrombosis of the subclavian-axillary vein occurring in the thoracic outlet in 1875.

B. Dr. William Halsted was one of the pioneers of modern surgical technique in the United States, using aseptic technique and novel wound closure techniques to advance the safety and efficacy of surgery. He performed the first emergency blood transfusion, the first radical mastectomy, the first nerve block, the first inguinal hernia repair, and invented the surgical glove. Dr. Halsted was named the first surgeon-in-chief of the Johns Hopkins Hospital, and was the first professor of surgery at Johns Hopkins Medical School. Dr. Halsted published a number of papers providing descriptions of subclavian artery aneurysms caused by cervical ribs in the late 1910s.

C. Dr. Alfred Washington Adson created and headed the Section of Neurological Surgery at the Mayo Clinic. He was a pioneer in American surgery, and was a founding member and president of the Society of Neurological Surgeons. Adson and Coffey first suggested the mechanism of the anterior scalene muscle causing upper extremity neurovascular compression in patients with cervical ribs in 1927.

D. Dr. Howard Christian Naffziger, who trained under eminent surgeons William Halsted and Harvey Cushing, became one of the most esteemed neurosurgeons of his time. He created the division of Neurosurgery and served as Chairman of the Department of Surgery at University of California San Francisco, was elected president of the American College of Surgeons, and was Chairman of the committee that established the American Board of Neurological Surgeons. Naffziger and Grant first introduced the concept of neurovascular compression in the thoracic outlet due to scalene muscle anomalies, without the presence of a cervical rib. They performed the first scalenotomies for relief of these symptoms in the 1930s.

E. Dr. Alton Ochsner was named Chairman of Surgery at Tulane Medical School at the young age of 31, and founded the world-famous Ochsner clinic at Charity Hospital in New Orleans, which remains one of the pre-eminent surgical teaching programs in the country. Dr. Ochsner was the first to report the link between cigarette smoking and lung cancer, and he trained some of the most prominent surgeons of the time, including Dr. Michael DeBakey. Dr. DeBakey is one of the most renowned cardiovascular surgeons in the world. He created the concept that became the Mobile Army Surgical Hospital (M*A*S*H unit) that had stellar success during the Korean War. Dr. DeBakey was one of the first cardiothoracic surgeons to perform coronary bypass surgery, was the first man to perform carotid endarterectomy, and made numerous
other contributions and innovations in cardiovascular surgery, including work on the Dacron artificial graft, the heart-lung machine, and the artificial heart. Drs. Ochsner, DeBakey, and Mims Gage at LSU published a comprehensive study of patients with symptoms of neurovascular compression in the thoracic outlet in the absence of a cervical rib in 1935, for which they coined the term, “Scalenus Anticus Syndrome”.

Our understanding of thoracic outlet syndrome has rapidly grown over the past several decades. In the last 50 years, over 1700 peer-reviewed journal articles regarding thoracic outlet syndrome have been published, with over 450 peer-reviewed articles published in the last 10 years alone. Medical researchers have published their extensive experiences evaluating and treating large numbers of patients with thoracic outlet syndrome at prestigious institutions such as Johns Hopkins University, Harvard Medical School, Yale University, University of Pennsylvania, New York University, Northwestern University Medical School, Washington University, University of California San Francisco, University of Colorado, University of Washington, Thomas Jefferson University, The Mayo Clinic, University of Michigan, University of Texas Southwestern Medical School, Baylor University, Tulane University, Louisiana State University and University of California Los Angeles(1-45). Many major university and teaching hospitals have developed dedicated clinics for the diagnosis and treatment of thoracic outlet syndrome, including Stanford University, University of Washington, University of Tennessee, University of Illinois Medical Center, Strong Memorial Hospital, Columbia University, Washington University, Emory University, University of Alabama, Cornell University, University of Minnesota, University of Pittsburgh, University of Wisconsin, Ohio State University, University of Southern California, University of California Los Angeles, and Chicago University. TOS is also discussed on important medical websites available to the public and to medical practitioners, including WebMD/eMedicine, The National Institute of Neurological Disorders and Stroke, The National Pain Foundation, MedicineNet, MDConsult, The Spinal Injury Foundation and MedScape.

Thoracic outlet syndrome is widely recognized, diagnosed and treated at top medical centers throughout the United States, and patients with thoracic outlet syndrome are likely to be seen commonly in the routine clinical practice of neurologists, vascular surgeons, neurosurgeons, orthopedic surgeons, internists and physiatrists.

Anatomy and Pathophysiology

The neurovascular bundle from the neck and chest, including the brachial plexus, the subclavian artery, and the subclavian vein, passes through three anatomic compartments to reach the upper extremity on each side (Figure 1, Page 4). From medial to lateral, the brachial plexus and subclavian artery pass through the scalene triangle (bounded by the anterior and middle scalene muscles), the costoclavicular interval (bounded by the clavicle and the first rib), and the retropectoralis space (bounded by the posterior margin of the pectoralis minor muscle and the anterior chest wall). The subclavian vein takes a slightly different course, first passing anterior to the anterior scalene muscle, then joining
the brachial plexus and subclavian artery to pass through the costoclavicular interval and retropectoralis space(5, 46-52).

Each of these three compartments may be narrowed by a number of processes that result in compression of the vital structures that pass through them (Figure 2, Page 5). The scalene triangle may be narrowed by abnormal size, tension, origins or insertions of the scalene muscles. The costoclavicular interval may be narrowed by congenital bony anomalies of the vertebrae, clavicle, or ribs, by healed fractures of the clavicle or ribs, or by abnormal positions of the shoulder girdle or clavicle. The retropectoralis space may be narrowed by abnormal positions of the scapula or clavicle, or by enlargement of the pectoralis minor muscle. These processes may cause isolated compression or entrapment of the brachial plexus(3, 23, 26, 53-56), the subclavian artery, or the subclavian vein. However, there is most often predominant compression of one structure, with lesser degrees of compression of the other structures.
Figure 2: Representative Pathologic Changes in Thoracic Outlet Syndrome

The processes that narrow the anatomic compartments may be classified as structural, functional, or post-traumatic. Structural processes include congenital muscle anomalies, fibrous bands and variant ligaments at the thoracic outlet, variant courses of the brachial plexus components, and bony abnormalities such as the cervical rib. These have been extensively documented and categorized by surgeons, anatomists and pathologists (7, 10-12, 19, 20, 23, 28, 38, 42-44, 53, 57-64). These anomalies, bands and ligaments, or the less common bony abnormalities, decrease the baseline dimensions of the three anatomic compartments. Narrowing of any of these compartments at baseline predisposes the patient to nerve or vascular compression after an episode of trauma, or after overuse or non-physiologic use of the upper extremities.

Functional abnormalities include overuse or non-physiologic use of the muscles of the shoulder girdle in occupational or recreational settings, resulting in hypertrophy of the scalene muscles or imbalance of the extensive shoulder girdle musculature. These functional abnormalities may result in direct narrowing of the anatomic compartments by the hypertrophied muscles, or in secondary narrowing of the anatomic compartments through abnormal positions of the clavicle and shoulder girdle relative to the chest wall.
It should also be noted that the position of the shoulder girdle is highly dynamic, and that the clavicle undergoes a complex three-dimensional motion with flexion or abduction at the shoulder joint. In particular, the clavicle moves to the greatest extent in a posterior direction, to a lesser extent in a superior direction, and rotates along its long axis. Since the first rib is relatively static, clavicular motion on flexion or abduction of the shoulder narrows the costoclavicular interval.

Post-traumatic abnormalities include direct trauma or stretching of the brachial plexus in motor vehicle accidents, fractures of the clavicle or ribs, and soft tissue injuries of the scalene muscles or other supporting structures of the neck (10-12, 19, 20, 28, 38, 42-44, 53, 57, 59-61, 65, 66).

Any congenital anatomic abnormality, functional change in the shoulder girdle, or post-traumatic alteration of the soft tissue or bony structures in the thoracic outlet can predispose the patient to compression of the brachial plexus as it passes through the thoracic outlet, especially with use of the affected upper extremity.

Thoracic outlet syndrome is divided clinically into three forms, based on which of the vital structures is compressed:

1. **Neurogenic TOS**-compression of the brachial plexus.
2. **Arterial TOS**-compression of the subclavian or axillary arteries.
3. **Venous TOS**-compression of the subclavian or axillary veins.

Neurogenic TOS is by far the most common form of TOS, accounting for 95 to 98% of all TOS cases. Unfortunately, this form of TOS has the most confusing clinical presentation of all the forms, and it is the form most likely to present in a subtle and insidious manner, and to follow a chronic and progressive course. No "gold standard" test has been accepted by the medical community for the diagnosis of neurogenic TOS.

Arterial and venous TOS are very uncommon, accounting for 5% or less of cases of TOS. These forms of TOS have dramatic clinical presentations, and are readily and accurately diagnosed with objective vascular imaging tests. Arterial TOS typically presents with upper extremity arterial insufficiency symptoms or embolic episodes, and ultrasound or arteriography demonstrates subclavian artery aneurysm, arterial thrombosis, or distal emboli. Venous TOS (Paget-Schroetter syndrome) typically presents with arm swelling and cyanosis, and ultrasound demonstrates thrombosis in the subclavian-axillary venous system.

The remainder of this white paper focuses on neurogenic TOS, because of its anatomic complexity, its diagnostic dilemma, and the past history of controversy regarding neurogenic TOS.
Clinical Presentation

Neurogenic thoracic outlet syndrome is a chronic compressive and entrapment neuropathy of the brachial plexus. Chronic nerve compression in rodents and primates has been well studied, and these histopathologic changes occur as a predictable and progressive continuum of changes. The clinical changes seen in neurogenic thoracic outlet syndrome parallel these histopathologic changes.

Initial disturbance of intraneurral blood flow leads to early breakdown of the blood-nerve barrier, which is followed by edema within the affected nerve fascicles. Fibrosis and thickening of connective tissue structures within and around the fascicles then develops. With continued compression, segmental demyelination occurs, eventually progressing to diffuse demyelination. Finally, the underlying axons undergo Wallerian degeneration. The degree and rapidity of these changes is related to the degree and duration of nerve compression, and within any one nerve, certain fascicles are affected more than others. Thus, sensory and motor symptoms and signs may take different courses.

The clinical symptoms and signs of neurogenic TOS parallel this temporal pattern. Initial sensory nerve compression occurs only with specific postures or maneuvers, resulting in pain, paresthesias, or vascular disturbances of the neck, shoulder, and upper extremity. These symptoms often occur hours after the compressive episodes, often waking the patient at night or being noted by the patient after waking from sleep. The patient may be completely asymptomatic at rest. In many cases, the patient learns to avoid these postures or maneuvers, either limiting or stabilizing the symptoms. If the patient is unable to limit or avoid these postures or maneuvers, the sensory symptoms may become persistent, even after the patient returns to a normal posture. Without resolution of the causative compression, the sensory symptoms can progress to anesthesia in some or all of the affected areas.

Initial motor nerve compression causes poorly localized pain or a cramping sensation in the muscles of the upper extremity. If the nerve compression persists, damage to the larger and more centrally located motor axons will occur, and weakness will gradually appear. Finally, in the most severe and persistent nerve compression syndromes, the large motor axons will degenerate, neurotrophic changes in the subtended muscles will occur, and atrophy of these muscles will be clinically detectable. These motor changes are the first objective clinical signs of TOS, but they occur late in the course of the disease process, and often represent permanent and irreversible nerve damage, with accompanying functional disability.

Thoracic outlet syndrome is well-known to occur in patients with cervical spine injuries following motor vehicle accidents. It is also common in those who use a computer keyboard or mouse for extended periods of time, such as office workers, those working in the legal profession, and sonographers, or in those who must keep their arms and neck in unusual postures for extended periods of time, such as musicians. The widespread and growing use of computers over the last two decades has dramatically increased the
incidence of repetitive stress injuries, and TOS is being recognized with increasingly greater frequency.

**Diagnosis**

Several clinical maneuvers and tests are used in patients with thoracic outlet syndrome, including Adson’s maneuver, the Wright test, and the Halsted maneuver. Positive test results are due to compression of the arterial system, rather than to compression of the brachial plexus. Sensitivity and specificity of these tests for compression of the brachial plexus is limited (67-69).

Roos’ test reproduces the patient’s symptoms when the patient’s arms are maintained in a position of neural tension and/or compression. The sensitivity and specificity of this test is not verified to date, and the test does not isolate or demonstrate a specific point of nerve compression.

Electromyography and nerve conduction velocity (EMG/NCV) studies have limited utility in the diagnosis of thoracic outlet syndrome. These examinations are limited by the difficulties in placing electrodes over the proximal components of the brachial plexus, by the inability to measure the length of these proximal components to determine velocity, by the small area of nerve damage relative to the length of the nerve, and by the pathophysiology of chronic nerve compression. In chronic nerve compression, smaller sensory nerves are affected earlier and to a greater extent than are the larger motor nerves, and there is a long continuum of progressive nerve damage, from segmental demyelination to complete demyelination to Wallerian degeneration of the axons. There is controversy regarding the point in this continuum at which EMG/NCV will detect an abnormality. Interestingly, there is a small amount of evidence demonstrating abnormal function of the proximal brachial plexus at surgery (38), but these areas are inaccessible to routine clinical EMG/NCV. When EMG/NCV abnormalities are present in patients with TOS, advanced nerve damage is present (due to fibrous bands in the thoracic outlet), with motor abnormalities that are very unlikely to resolve after surgery (38, 70). Therefore, EMG/NCV is used primarily to rule out other peripheral neuropathies, rather than to rule in TOS.

Cervical spine radiographs are frequently utilized in the evaluation of patients with thoracic outlet syndrome, as early descriptions of the syndrome invariably involved patients with cervical ribs. However, as knowledge of TOS progressed, it became apparent that the vast majority of cases of neurogenic TOS are associated with soft tissue abnormalities, without cervical ribs. Currently, cervical spine radiographs have a limited role in the evaluation of these patients. If cervical ribs are present in a patient with the typical presentation of TOS, the diagnosis is easily made. Most of these patients have the arterial form of TOS. If cervical ribs are not present in a patient with the typical presentation of TOS, the diagnosis is vastly more challenging.
Imaging of Thoracic Outlet Syndrome

The diagnosis of TOS in patients with the typical clinical presentation relies on the demonstration of the predisposing soft tissue abnormalities or compression of the brachial plexus, subclavian artery or subclavian vein in the thoracic outlet. As outlined above, this is exceedingly difficult in the clinical evaluation of these patients. Unfortunately, at surgery, up to one third of patients have no predisposing anatomic abnormalities (43). In the absence of such abnormalities, the success rate of surgical decompression is low (53).

Therefore, accurate and reliable imaging of the anatomy of the thoracic outlet, visualization of the predisposing anatomic abnormalities, and demonstration of the compression of nerves, arteries, and veins is paramount for the accurate diagnosis of these patients.

Many studies have been published regarding the effectiveness of ultrasound (71-76), CT scanning (68, 77-85) or MRI scanning (46, 47, 86-103) in the evaluation of thoracic outlet anatomy, and in the diagnosis of TOS.

Ultrasound is excellent for the evaluation of blood flow in the arteries and veins of the thoracic outlet, produces no ionizing radiation, and can measure dynamic changes or compression of the blood vessels in real time. Additionally, it can be used to evaluate these blood vessels with the patient's upper extremities maintained in any position. A few reports have demonstrated the use of ultrasound in evaluating the anatomy of the thoracic outlet and the brachial plexus, but ultrasound is unable to image through bony structures, and the position of the clavicle causes significant limitation of its use in patients with TOS. Since the vast majority of cases of TOS are neurogenic, ultrasound is not suitable at present as a first-line diagnostic tool.

CT scanning is excellent for evaluation of the bony structures of the thoracic outlet, and can be a very rapid scanning technique. However, CT scanning utilizes ionizing radiation, requires iodinated contrast material for the evaluation of blood vessels, and does not differentiate soft tissues as well as MRI. Since many cases of TOS are due to soft tissue abnormalities rather than to bony abnormalities, CT scanning is likely to lack sensitivity and specificity.

MRI scanning is excellent for demonstrating soft tissue detail including the muscles, nerves, blood vessels and fatty areas of the thoracic outlet, produces no ionizing radiation, utilizes gadolinium contrast material for the evaluation of blood vessels (which has considerably fewer adverse effects than does iodinated contrast material), and creates images in multiple planes. However, MRI scanning requires more time than does CT scanning, and bony structures are less obvious on MRI than they are on CT. In experienced hands, MRI defines all of the vital structures of the thoracic outlet, and is the best modality for ruling out spinal stenosis and neural foraminal stenosis, which are important in the differential diagnosis in these patients. The medical literature on MRI continues to expand. Most recently, a review article describing the use of MRI for the
diagnosis of thoracic outlet syndrome has been approved for Continuing Medical Education by the Radiologic Society of North America, the largest professional organization of radiologists in the world (104).

Treatment

The appropriate treatment of patients with thoracic outlet syndrome depends on the clinical form of thoracic outlet syndrome, and the underlying anatomic cause of the syndrome.

Patients with venous thoracic outlet syndrome usually present with upper extremity thrombosis, which causes arm swelling, cyanosis, and visible collateral vessels. Rarely, these patients can present with a pulmonary embolism. Urgent thrombolysis is usually initiated, after which definitive treatment is performed, including either angioplasty of the vein, or surgical decompression of the thoracic outlet, with removal of the structures that are causing the extrinsic compression of this vein.

Patients with arterial thoracic outlet syndrome usually present with upper extremity pallor, coldness and distal emboli to the hand and/or fingers. Urgent thrombolysis is usually initiated, after which definitive treatment is performed, including surgical decompression of the thoracic outlet, with removal of the structures that are causing the extrinsic compression of the artery, as well as repair or bypass of the damaged artery, if necessary. Cervical ribs are much more common in this subtype of thoracic outlet syndrome, and are usually resected.

Patients with neurogenic thoracic outlet syndrome usually present with an insidious and progressive course, as described above. These patients almost always undergo a specialized and focused program of physical therapy for a period of weeks to months, which is aimed at rebalancing the muscles of the shoulder girdle and improving the patient's posture. Patients with repetitive stress injury may also undergo occupational therapy and ergonomic evaluation of their work environment. A small number of patients fail to respond to conservative therapy, at which point surgical decompression of the thoracic outlet is considered. It should be noted that patients with long-standing disease, as indicated by muscle atrophy or weakness in the hands, should undergo surgical decompression as soon as reasonably possible, as muscle atrophy or weakness indicates advanced and likely permanent nerve damage.

Surgical outcomes have been published from numerous university and private practice sites, both in the United States and abroad (4, 8, 14, 22, 27, 63, 105-113). Following surgical decompression of the thoracic outlet, 80 to 85% of patients report excellent or good results. During the first two postoperative years, some of these patients experience recurrent symptoms. However, beyond the first two postoperative years, approximately 70% of patients report excellent or good results. It should be noted that patients with long-standing symptoms and/or motor signs are less likely to experience symptomatic improvement or to regain motor function (38, 114). These findings closely parallel those
seen in other nerve decompression procedures, including neural foraminotomy, carpal tunnel release, and decompression of the ulnar nerve in the cubital tunnel (114).

Our Expertise

Dr. Scott Werden has spent the last several years developing and refining our patent-pending MRI scan for the evaluation of patients with thoracic outlet syndrome. He has personally supervised and interpreted almost 500 of these studies to date. He has consulted on a regular basis with local physicians who diagnose and treat patients with thoracic outlet syndrome, has observed surgery to review and confirm findings of MRI scans, and has been named as a retained expert on thoracic outlet syndrome in numerous medicolegal cases. Dr. Werden has been invited to speak on thoracic outlet syndrome at national meetings and at Grand Rounds presentations of local hospitals and universities. Dr. Werden has one of the largest databases on MRI scanning of patients with thoracic outlet syndrome, and he is in the process of publishing several papers on this topic in peer-reviewed journals.

27. Sanders RJ. Results of the surgical treatment for thoracic outlet syndrome. Semin Thorac Cardiovasc Surg 1996; 8:221-228.


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