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Etiology of neurogenic thoracic outlet syndrome

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Neurogenic thoracic outlet syndrome (TOS) occurs because there is insufficient space for the nervous structures. There is usually a congenital predisposition to develop TOS symptoms. An injury that may seem minor usually is superimposed on the congenital anomalies and compromises the space further.

The symptom complexes that are recognized as TOS have been known for more than 100 years. They are variously overdiagnosed or underdiagnosed, depending on one's perspective [1,2]. They are interesting syndromes because of their variations and the emotional discussion that surrounds them. The emotional aspect should be set aside and the clinical syndromes recognized. Having the syndrome does not necessarily lead to disability or a surgical procedure. As with most medical problems, there is a broad spectrum from mild to severe symptoms. In its mildest form TOS may be simply positional numbness of no clinical significance.

There is an anatomic basis for the symptom complex in patients who have this syndrome. For treatment to be effective, whether it is surgical or nonsurgical, the anatomic etiology of the symptoms must be understood. Without understanding the underlying anatomic variations in the thoracic outlet that adversely affect the brachial plexus and subclavian vessels, a patient's complex presentation becomes a conundrum of vague symptoms inexplicable by a routine physical examination. The understanding of anatomic aspects has evolved over time and continues to do so. With the advent of more detailed imaging techniques, cross specialty discussions are even more important in providing advances in understanding.

Terminology

Even the name "thoracic outlet syndrome" is shrouded in contentious controversy much to the detriment of the patients affected. It was first used by Peet in 1956 [3] and was defined as neurovascular symptoms of the upper extremity presumably caused by mechanical changes with narrowing of the spaces in the thoracic outlet through which the major nerves and vessels pass. The term was an attempt to unite the many other names the syndrome has been called into one term. Other names used, such as scalenus anticus syndrome, costoclavicular syndrome, cervical rib syndrome, subcoracoid-pectoralis minor syndrome, costoclavicular syndrome, and first-thoracic rib syndrome further confused the understanding of the pathophysiology of this condition. Whether it is the thoracic outlet or thoracic inlet is even disputed. Clinicians tend to call it the thoracic outlet because the structures being compressed are exiting the chest in this location. Anatomists consider this incorrect terminology, as they work from superior to inferior, and thus consider the same area to be the thoracic inlet [4]. These controversies over semantics only add further confusion to already complex clinical problems.

Understanding of the cause of TOS began with the recognition of cervical ribs and their role in arterial compression. Galen and Vesalius described cervical ribs in their dissections of human bodies in the middle ages. A 3000-year-old

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Egyptian mummy in a British museum was found on radiograph to have cervical ribs of uncertain clinical significance by the senior author. Arterial and venous problems were attributed to cervical ribs even before radiographs were available to identify them [5]. Later it became apparent that the vascular compressions could occur in the absence of cervical ribs and that the band connecting the cervical rib or elongated transverse process to the first rib was often the culprit rather than the rib itself. Once it was clear that congenital anomalies could compress nerves and vessels, it was realized that most of the symptoms experienced by these patients were not vascular, but were in fact neurologic. More anatomic anomalies and abnormal bands were described, and most of these affected nerves rather than vessels. As time passed, the differences between upper and lower plexus TOS were clearly defined, first by Swank et al in 1944 [6] and then by Roos [7]. Surgeons operating on these patients came to recognize the many anatomic abnormalities, but many neurologists recognized only the band that extends from the first rib or elongated transverse process when it caused atrophy in the hand. At that advanced stage the diagnosis is clinically obvious and is associated with abnormal electrophysiologic studies. One would hope that the diagnosis could be made before such permanent damage takes place [8,9].

Definition

For the purposes of definition, the authors consider the thoracic outlet to extend from the outer edge of the first rib laterally and to include the mediastinum medially. It continues upward in the neck to the level of the fifth cervical nerve root. This space contains the anterior and middle scalene muscles, the five primary cervical nerves of the brachial plexus, the three trunks, the phrenic nerve, long thoracic, suprascapular, and dorsal scapular nerves, the stellate ganglion, the subclavian artery and vein, the thoracic duct, scalene lymph nodes, and the apex of the lung. Although these structures are well described in anatomy books, clearly there is wide variability in their exact location. Technically the cervical nerve "root" consists of that portion of each nerve arising from the spinal cord and lying in the subdural space. Once it passes through the dura it is no longer a "root" but is then labeled a "cervical nerve".

Anatomic variations

TOS is a space problem. Occasionally neurovascular compression in the thoracic outlet arises from bony abnormalities that are either congenital or post-traumatic. Cervical ribs may be present or the C7 transverse process may be elongated. The C7 transverse process is considered "elongated" (and therefore to possess a taut type 2 band) if it extends beyond the tip of the T1 process immediately below it, as seen on cervical spine films. First rib deformities, exuberant fracture callus, or displaced fractures also may cause the problem. Rib hemangiomas [10], subluxated clavicles, and pseudarthroses [11] also have been described. An occasional patient has underlying fibrosis secondary to an injury or a local hemorrhage that produces scarring without the underlying anatomic abnormality. Patients who have recurrent TOS fit into this category, presuming that the first operation was done correctly. More commonly, patients with TOS have underlying congenital anomalies that *predispose* them to develop space problems, especially if trauma is superimposed. Many of these have been characterized and others are yet to be described. The careful surgeon actively seeks these abnormalities and deals with them specifically. Absent an anatomic explanation for the symptoms at the time of surgery, surgical attempts at decompression are generally unsuccessful.

The abnormalities found in patients with lower trunk brachial plexus problems (C8–T1) are shown in Fig. 1.

Patients presenting with upper plexus neurologic symptoms (arising from C5, C6, and C7) have different anomalies from those with lower plexus symptoms. The anomalies contributing to upper plexus symptoms are shown in Fig. 2.

Swank characterized the anomalies leading to upper brachial plexus symptoms somewhat differently [6], illustrating the complexity of the anatomic situation found at surgery on these patients.

- Hypertrophy of the anterior scalene in all patients
- Origin of the anterior and middle scalene muscles from a common belly with low division of the two, so that the brachial plexus was compressed between the subclavian artery and the fusion of the two muscles
- Passage of the brachial plexus through the substance of the scalenus anticus muscle

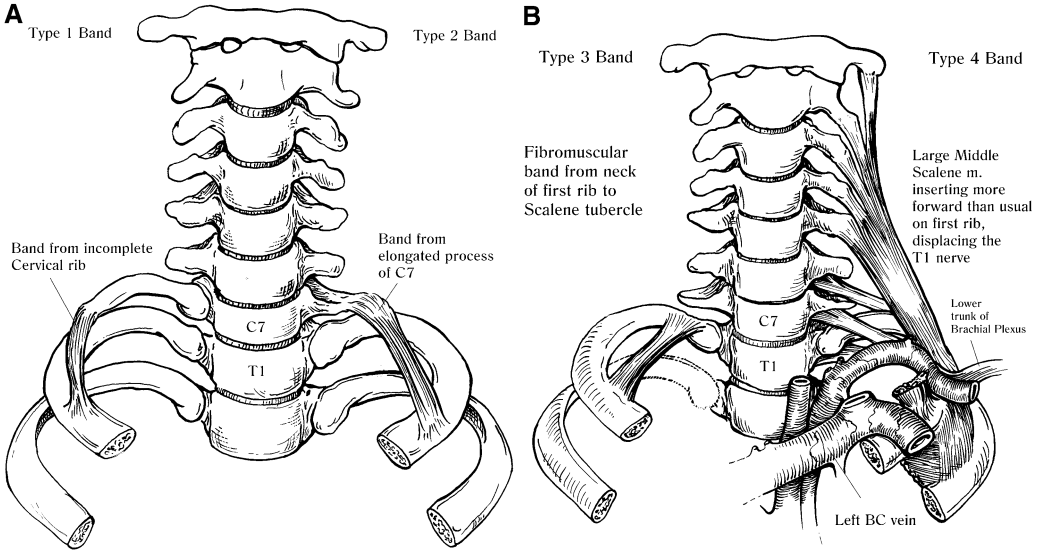

Fig. 1. Fibrous bands and congenital anomalies affecting vessels and lower trunk of the brachial plexus. (*A*) Type 1: A tight fibrous band from the tip of an incomplete cervical rib to the mid portion of the first rib, passing beneath the T1 nerve of the brachial plexus (*left*). Type 2: A tight fibrous band, similar to Type 1, but originating on an elongated C7 transverse process. This transverse process on x-ray may be seen extending more laterally than the T1 transverse process. The band usually attaches to the first rib just posterior to the scalene tubercle and likely represents an abortive attempt to form a cervical rib (*right*). (*B*) Type 3: An extra fibro-muscular band arising from the neck of the first rib medially passing anteriorly under the T1 nerve and attaching to the inner aspect of the first rib behind the scalene tubercle (*left*). Type 4: A large middle scalene muscle inserting more forward on the first rib than normal and displacing the T1 nerve. The nerve thus deviates anteriorly around the anterior surface of the middle scalene muscle. Often this muscle has a fibrous, sharp anterior edge, which abuts against the T1 nerve. Type 4 causes pain over the back of the shoulder radiating through the triceps area and down the inner arm to the ring and small fingers. A variant of this anomaly, classified as Type 4-B, is actually a white fibrous band that pins the T1 nerve against the vertebral body (*right*). (*C*) Type 5: Scalenus minimus muscle. This muscle arises from the C6 and C7 transverse processes and passes parallel to the anterior scalene muscle but lies behind the artery instead of in front of it and attaches posteriorly on the first rib. The scalenus minimus is located between the nerves and the subclavian artery (*left*). Type 6: A scalenus minimus muscle, which attaches to Sibson's fascia, the reflection of the endo-thoracic fascia covering the cupola of the lung. This band remains intact after a complete removal of the first rib so it must be recognized and dealt with separately (*right*). (*D*) Type 7: A long fibro-muscular band stretching from the lower part of the middle scalene muscle passing anteriorly under T1, the subclavian artery and the vein to attach to the costal cartilage or sternum (*left*). Type 8: A fibro-muscular band arising from the anterior scalene muscle and passing under the subclavian vein to attach to subclavius muscle and costal cartilage. This band is a main cause of subclavian vein thrombosis (*right*). (*E*) Type 9: A sharp edged fibro-muscular band attached along the posterior inner surface of the first rib lying tightly against T1 nerve. This can sometimes be seen to indent the nerve (*left*). Type 10: A double fibrous band, which forms a "V" over the cupola of the lung. The posterior limb arises from the cervical rib or neck of the first rib attaching to the middle third of the first-rib. The anterior limb arises from the point of attachment of the posterior limb and passes under the vein to the sternum or costal cartilage (*right*). (*Modified from* Roos DB, Annest SJ, Brantigan CO. Historical and anatomic perspectives on thoracic outlet syndrome. Chest Surg Clin N Am 1999;9:713–23; with permission.)

- Compression of the nerves of the brachial plexus by the scalenus anticus muscle against the scalenus medius muscle, which was held forward by a transverse process of the seventh cervical vertebra or compression by cervical ribs
- Abnormally broad insertion of the scalenus anticus muscle to the scalene tubercle of the first rib

However one characterizes the anomalies of the scalene muscles, they must be sought and understood at surgery. Each set of abnormalities presents unique hazards to the unwary surgeon.

The abnormality found in patients with venous TOS is consistently present in these patients. The anterior scalene muscle has a band of muscle extending under the vein like a sling connecting to

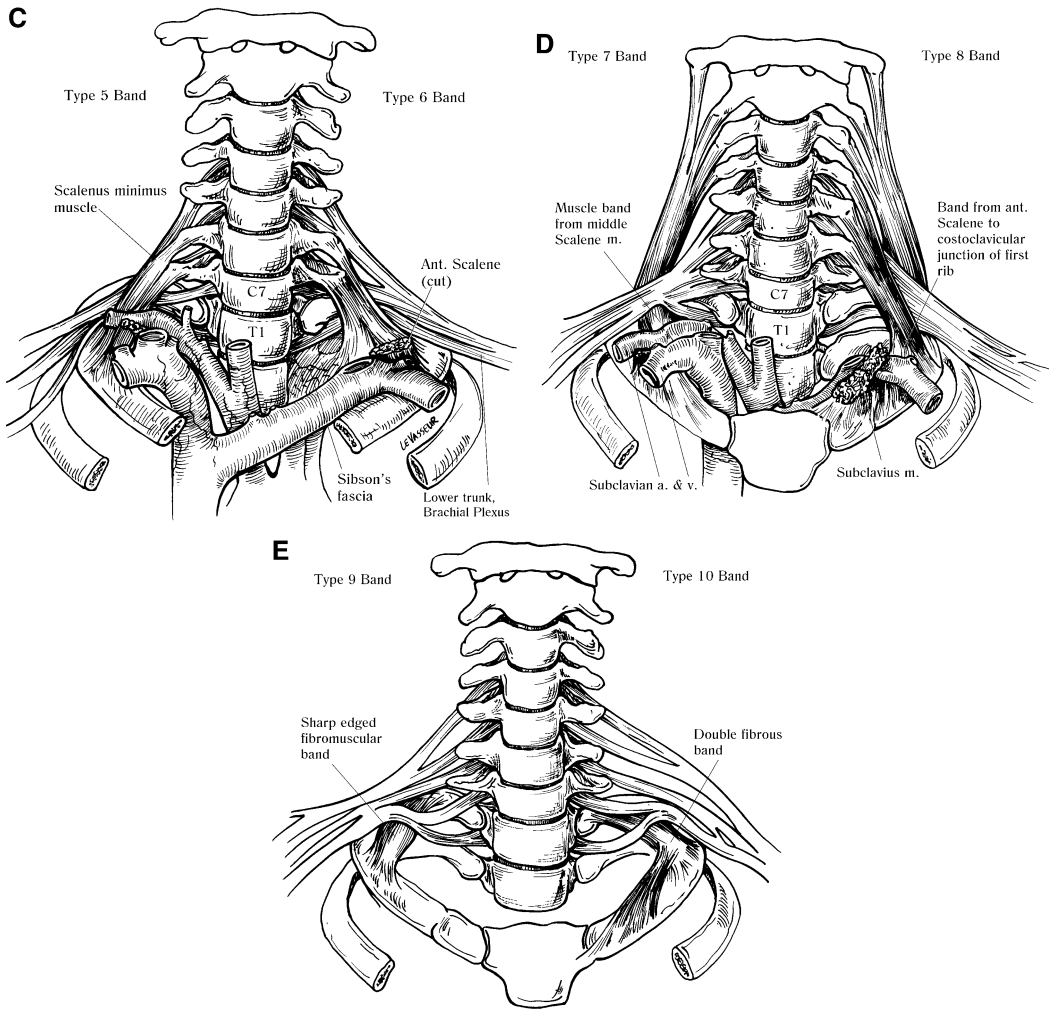

Fig. 1 (*continued*)

the subclavius muscle. This catches the subclavian vein in a scissor mechanism.

Trauma

Given the anatomic predisposition to develop TOS, there often is a superimposed injury that precipitates the symptoms of TOS. This may be a simple exercise-induced hypertrophy of the scalene muscles or sagging of the shoulders with age and poor posture. Typical specific injuries include whiplash injuries [12], blows to the shoulders, and repetitive trauma. These injuries produce cervical muscle spasm that puts traction on the abnormal structures of the thoracic outlet. Spasm leads to swelling of the muscle. Traction leads to edema and swelling of the nerves. Swelling and spasm further aggravate the space problem, leading to more compromise. As the pathophysiology becomes established, scarring and fibrosis add to the problem. These injuries set up a vicious cycle. Pain, bad posture, poor physical conditioning, and anxiety then aggravate that cycle.

Controversy

Many neurologists consider a patient to have TOS only if there is muscle wasting in the hand together with abnormal electrophysiologic studies. Some speak in terms of "surgically proven" TOS in patients with a band extending from a cervical rib or a long transverse process elevating and irritating the lower trunk of the brachial plexus associated with atrophy. The

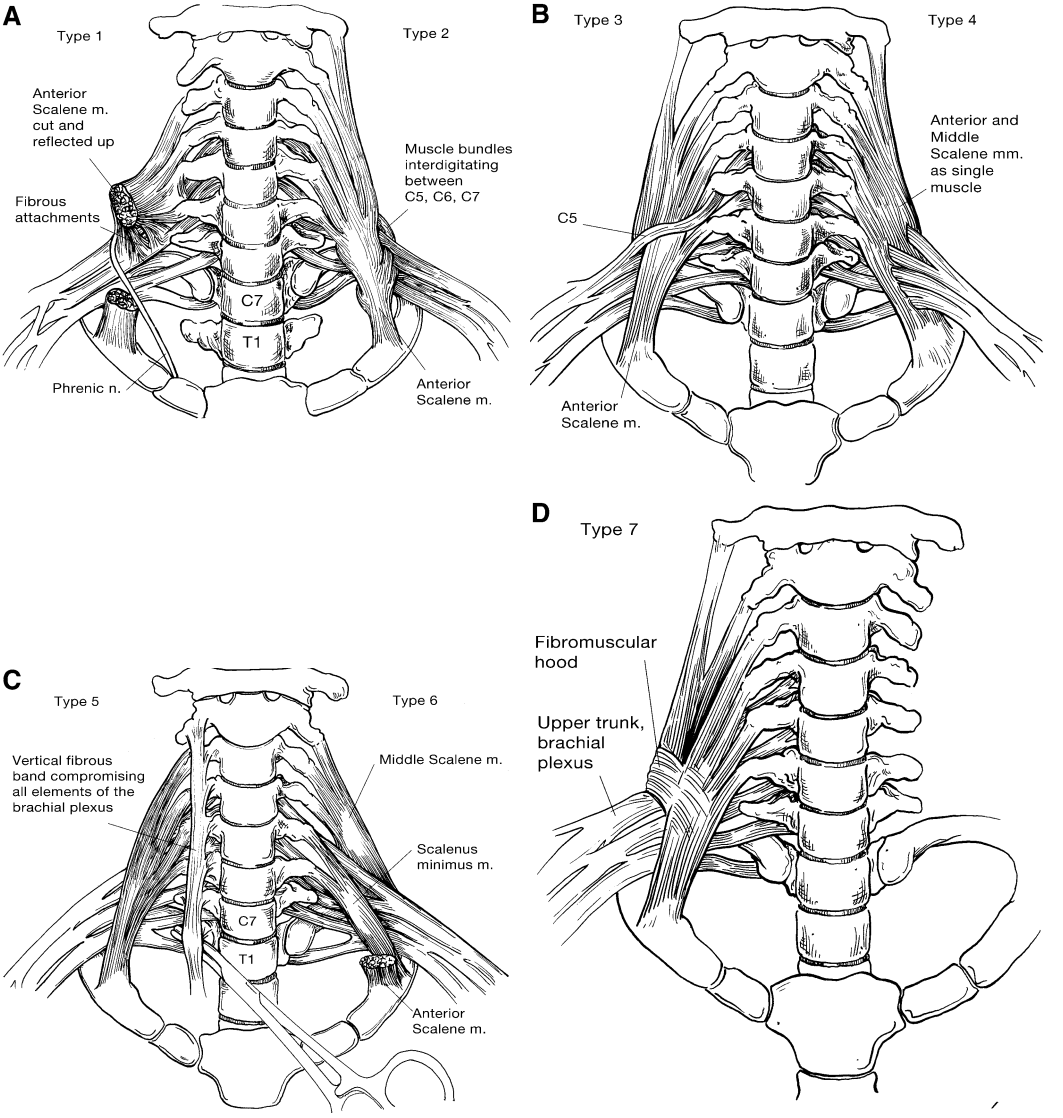

Fig. 2. Congenital anomalies affecting the upper and middle trunks of the brachial plexus (*A*) Type 1: Anterior scalene muscles fused to or seeming to grow into the epineurium of the C5 and C6 cervical nerves (*left*). Type 2: Bundles of the anterior scalene muscle that connect with the middle scalene muscle and interdigitate between C5, C6 and C7 nerves (*right*). (*B*) Type 3: Anterior scalene muscle that lies between C5 and C6 and becomes continuous posteriorly to C5 with the upper portion of the middle scalene muscle. The C5 nerve and sometime even the upper trunk of the brachial plexus are displaced so they lie anterior to the anterior scalene muscle (*left*). Type 4: The scalene muscles presenting as a single mass of muscle without division into separate anterior and middle bellies. The individual nerves penetrate this mass like arrows (*right*). (*C*) Type 5: Separate fibro-muscular bands that cross between C7 and C8 medial to and deeper than the anterior scalene muscle. These may be difficult to see but can frequently be palpated and feel like violin strings (*left*). Type 6: C7 and C8 nerves covered with a separate muscle distinct from the anterior scalene muscle. This is likely the origin of the scalenus minimum muscle (*right*). (*D*) Type 7: A fibro-muscular hood arching over the proximal aspect of C5 connecting the anterior and middle scalene muscles. This hood cinches down over the superior edge of the C5 nerve during scalenus contraction compressing the C5 nerve and upper trunk. (*Modified from* Roos DB, Annest SJ, Brantigan CO. Historical and anatomic perspectives on thoracic outlet syndrome. Chest Surg Clin N Am 1999;9:713–23; with permission.)

other bands and anomalies described previously are "surgically proven" also. Not believing that a condition exists because a test is negative is an example of the myopic approach taken by some physicians who do not believe it exists unless one can see it on a radiograph. Unfortunately there are many who believe the most common type of neurogenic TOS does not even exist because they cannot see or measure it. That reasoning is akin to believing that the atom did not exist until the twentieth century or that the presence of palpable pulses rules out claudication.

Seeing is believing! The doubtful should take the time to attend a surgical procedure to release the compressed nerves in the outlet wherein the precise anatomic anomalies that cause the specific symptoms of each cervical nerve affected are readily visualized. Then follow the patient in the early postoperative period to appreciate the remarkable relief of prolonged distressing and often disabling symptoms most of those patients achieve almost immediately. Such a scenario would offer a simple resolution of the pervasive controversy surrounding this diagnosis and would lead to much more appropriate and compassionate medical or surgical care that has so often been denied.

Summary

The anatomic problems that lead to TOS are now well known. They consist of congenital anomalies that are superimposed on some form of trauma. There are some promising technologies that offer hope of early anatomic diagnosis. Sophisticated imaging of the brachial plexus as advocated by Collins offers hope [13]. High resolution multidetector CT scanning seems even more promising.

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