



Neurogenic TOS Comprehensive Overview

SYMPTOMS

Neurogenic TOS is when the brachial plexus nerves are compressed within the scalene triangle and/or the costoclavicular space. It is a nerve compression syndrome which is a condition wherein nerves are being compressed or pinched/squeezed by other internal body structures resulting in nerve damage or dysfunction leading to symptoms. Carpal tunnel syndrome is also a nerve compression syndrome. The difference is that, with carpal tunnel, one nerve is compressed lower down in the arm by the wrist, and with NTOS, there are several different nerves being compressed up near the neck and chest. Since the brachial plexus supplies the nerves to the chest, most of the upper back, shoulder, arm, and hand, symptoms can be experienced in any of these places. NTOS can cause several different types of symptoms including nerve symptoms, muscle symptoms, and even symptoms that affect blood flow.

Symptom Presentation:

- Majority of patients are female (approx. 60-70%)
- Most frequently presents in patients ages 20-40, but can & does occur outside of this range
- Usually presents on 1 side of the body at least initially, and typically the side of the body with the dominant arm is the side to first show symptoms
- tend to start gradually and worsen over time but sudden onset can also occur
- can vary throughout the day or from day-to-day in location, type, and severity especially early on in the condition
- tend to worsen during sleep with many patients having trouble falling asleep, waking up during the night due to symptoms, or having symptoms upon waking in the morning. Sleeping positions which can exacerbate symptoms are sleeping with the arm overhead, sleeping on the back, and sleeping on the affected side
- Can range from mild and mostly positional to constant severe and debilitating pain that can result in patients not being able to work, socialize or take care of themselves which can also have a severe impact on the mental health of the patient
- Some symptoms worsen with certain TOS-aggravating activity such as heavy lifting or heavy use of the arms, pushing or pulling too much weight, repetitive use of the arms, overhead use of the arms, keeping the arm in an outstretched or awkward position for prolonged periods of time, and anything that involves fine motor skills/fine finger movements. Such activity examples include:
 - o driving
 - o brushing teeth
 - o talking with the phone up to ear
 - o brushing, washing, blow drying, or styling hair
 - o typing/texting
 - o using a mouse
 - o vacuuming
 - o folding laundry
 - o sewing
 - o writing
 - o throwing
 - o playing a musical instrument such as guitar or violin

Common Symptoms:

- Pain in shoulder, arm and/or hand. Pain can be any type such as dull, sharp, shooting, stabbing, squeezing, aching, burning, electrical zaps
- Numbness or tingling in shoulder, arm and/or hand. Most commonly in the pinky and ring fingers, but it can involve any or all the fingers.
- Hypersensitivity of the skin of the shoulder, arm and/or hand
- Pain radiating down the arm to the hand particularly when turning the neck toward the affected side
- Feeling of fatigue or heaviness in the arm especially when elevated
- Feeling of hot or cold sensations within the shoulder, arm, and/or hand
- Temperature and color changes of the hand (cold & bluish purple) or (hot & red) especially when arm is elevated or hanging straight down (see below to learn more about the role of NTOS in these types of symptoms)
- Swelling of the hand and/or fingers
- Loss of hand dexterity such as inability to grip or pinch objects and easily dropping things
- Enlarged or bulging veins in the arm and/or hand
- Atrophy and weakness in the muscles of the fleshy base of the thumb (Gilliat-Sumner Hand)
- Claw hand (characterized by curved or bent fingers, making the hand appear claw-like)
- Muscle spasm and twitching in the arm and/or hand
- Swelling at the base of the neck just above the collarbone
- Neck muscle spasm, tension, tightness, and pain particularly on the sides and back of the neck including in the form of trigger points with specific pain patterns radiating to other parts of the body.
- Tenderness in the base of the front of the neck just above the collarbone that might also reproduce symptoms radiating down the arm
- Headaches in the back of the head (occipital headaches) which can wrap around the back of the head to above the eye.
- Collarbone pain
- Shoulder blade pain (often sharp or knife-like)
- Winged shoulder blade (shoulder blade is prominent & sticks out from the back instead of lying flat)
- Upper back muscle spasm, tightness, and pain particularly in upper trapezius, rhomboid, levator scapulae, and muscles that surround the shoulder blade
- Pain in the chest, breast and armpit can also go along with NTOS in the form of Pec Minor Syndrome. Go to www.tosoutreach.com to learn more about the symptoms of Pec Minor Syndrome
- Rarely, patients with NTOS will also have VTOS. Go to www.tosoutreach.com to learn more about the symptoms of VTOS.
- Rarely, patients with NTOS will also have ATOS. Go to www.tosoutreach.com to learn more about the symptoms of ATOS.

Less Common Symptoms:

- Pain, numbness or tingling in the jaw or face
- Migraine headaches (although no known connection to NTOS)
- Atrophy or wasting of the muscles in the upper back, shoulder, arm, and/or hand (aside from what is mentioned above)
- Decreased range of motion in the shoulder and arm
- Decreased range of motion in the neck (mainly due to muscle spasm)
- Ear pain or tinnitus (ringing or pulsing in the ear)

- Dizziness
- Fainting or passing out particularly with neck or head movement
- Shortness of breath (due to compression of the phrenic nerve)
- Heart palpitations or fast heart rate (due to compression of the vagus nerve)

Additional Information Related to the Role of NTOS in Causing Vascular Symptoms

Symptoms related to blood flow changes such as changes in temperature and color and swelling are actually quite common in patients with NTOS. These symptoms are often assumed to be caused by Arterial or Venous TOS, but typically, this is not the case. It's actually caused by a sympathetic induced vasoconstriction. The sympathetic nervous system controls the blood vessels including how constricted (closed down) or how dilated (opened up) they are. The brachial plexus nerves contain fibers by which they communicate with the sympathetic nervous system. Therefore, when the nerves are compressed, these fibers are compressed and can cause the sympathetic nervous system to become overactive. Being overactive means that it can tell the vessels to overconstrict which reduces blood flow resulting in coldness and/or blue/purple/mottled discoloration. It can also tell them to open too wide resulting in swelling, redness, bulging veins, etc. Arm elevation and arm hanging down positioning further irritates the nerves by either compressing them more or stretching them which can enhance these vasoconstriction symptoms.

DIAGNOSIS

DIAGNOSIS CHARACTERISTICS

The diagnosis of NTOS has long been quite elusive and even controversial, particularly to those health care providers who do not specialize in TOS. The reason is that there is no single test or imaging study that can definitively prove or disprove the existence of NTOS. It is a clinical diagnosis which means that the diagnosis relies heavily on the patient's symptoms, medical history, and physical examination. The clinical diagnosis of NTOS is then further supported by objective testing and imaging to rule out more common medical conditions that can mimic NTOS and is then supplemented with a few NTOS-specific clinical tests. It is also often referred to as a diagnosis of exclusion due to the fact that a large part of the diagnostic process is comprised of ruling out other more common conditions whose symptoms can mimic those of NTOS. Aside from objective testing, ruling out these other conditions often involves doctors who specialize in these conditions to assist with determining whether these conditions are causing or contributing to the patient's symptoms. The journey for most patients begins with a primary care physician and then often referrals to specialists such as orthopedic surgeons, neurologists, peripheral nerve surgeons, neurosurgeons, and physiatrists. Most of these referrals and specialists will not have NTOS in mind. For some, a chiropractor or physical therapist will be the first to mention TOS, and the patient will then begin getting referrals with the hope of getting a TOS diagnosis. For many, these referrals and specialist visits will not end with a diagnosis of NTOS. However, luckily, most of these patients will still make progress toward a diagnosis of NTOS through the testing and procedures ordered for them which will have almost certainly ruled out other more common medical conditions. Unfortunately, for some of them, this process will also include having had unsuccessful surgeries for conditions which are ultimately found to be most likely inaccurate diagnoses.

Who Diagnoses & Treats NTOS?

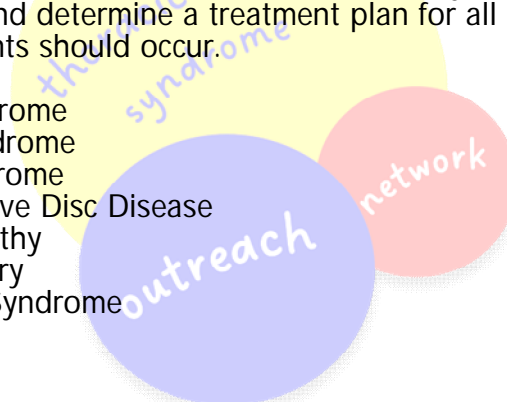
Any doctor who is familiar with NTOS can give a preliminary diagnosis. These most commonly are orthopedic surgeons, peripheral nerve surgeons, or neurologists. Once a

diagnosis of NTOS is suspected, referral should be made to a TOS specialist to make a definitive diagnosis. TOS specialists are usually vascular surgeons and occasionally cardiothoracic surgeons. However, most vascular surgeons and cardiothoracic surgeons DO NOT specialize in TOS. In fact, most of them have very little experience with TOS. Therefore, it needs to be a vascular surgeon or cardiothoracic surgeon who specifically specializes in TOS. Although it might seem a bit unconventional, these surgeons treat all three types of TOS including NTOS. There are some orthopedic surgeons, peripheral nerve surgeons, and neurosurgeons who treat NTOS, but those specialties are in the minority and are not generally represented within the community of high volume TOS specialists. However, outside the US, where TOS experience and knowledge is more limited, it is somewhat more common to see these other specialties represented in the options for TOS specialists. For help finding a TOS surgeon, go to www.tosoutreach.com/find-a-surgeon.

Conditions That Can Mimic NTOS:

Below is a list of medical conditions that have symptoms which can mimic the symptoms of NTOS. Many of these conditions are more common than NTOS, and thus should be definitively ruled out before any further steps toward a diagnosis of NTOS are taken. To make things even more complex, it is possible for NTOS to coexist with these other conditions meaning that a patient can have both NTOS and one or more of the conditions that can mimic it. An experienced TOS specialist in combination with specialists in these other conditions will likely be necessary in a scenario where a patient has been found to have both NTOS and another one of these conditions. Ideally, the specialists will collaborate to confirm all diagnoses and determine a treatment plan for all conditions particularly as to in what order the treatments should occur.

- Carpal Tunnel Syndrome
- Cubital Tunnel Syndrome
- Radial Tunnel Syndrome
- Cervical Degenerative Disc Disease
- Cervical Radiculopathy
- Brachial Plexus Injury
- Parsonage-Turner Syndrome
- Rotator Cuff Injury
- Bicep Tendonitis
- Cervical Dystonia
- Complex Regional Pain Syndrome (CRPS)
- Fibromyalgia
- Multiple Sclerosis
- Lymphedema
- Pancoast Tumor
- Raynaud's Syndrome
- Vasculitis
- ALS (Lou Gehrig's Disease)



TESTING

Radiology (X-Ray Imaging)

There is no radiology test that will definitively show NTOS. Therefore, most x-ray imaging would be done to rule out other conditions that mimic NTOS. Specific imaging tests would likely be ordered based on history and symptoms. The tests listed below will not all be performed and are not all required.

Plain Chest X-Ray can be done to look for any anatomical abnormalities such as anomalous first rib, rib fracture, or collarbone fracture.

Plain Cervical Spine (Neck) X-Ray can be done to look for cervical degenerative disc disease or to look for cervical ribs or elongated C7 transverse processes. Most radiologists are not looking for cervical ribs or elongated C7 transverse processes as they are typically incidental findings and most people who have them do not experience symptoms or develop any conditions related to them. Even if a radiologist does see them, they often leave them out of the radiology report for these same reasons. If a patient is having this x-ray specifically to help with diagnosis of NTOS, they should request that the ordering physician state that the purpose of the imaging is to rule out cervical ribs. However, the presence of cervical ribs alone is not diagnostic of NTOS. In fact, the majority of patients with NTOS do not have cervical ribs. Only around 10% or less of NTOS patients have them, and they are not required for diagnosis.

Plain Shoulder X-Ray can be done to look for any bony or joint abnormalities or to check for shoulder joint dislocation.

Cervical Spine (Neck) CT Scan mainly done to look for and rule out degenerative disc disease of the cervical spine. It can also help identify cervical ribs and elongated C7 transverse processes.

Cervical Spine (Neck) MRI mainly done to look for and rule out cervical nerve root compression and radiculopathy in addition to compression of the spinal cord usually in the form of disc herniations or neuroforaminal narrowing. It can also help look for any other soft tissue abnormalities around the cervical spine and nerve roots as well as help identify cervical ribs although not as good as cervical x-ray or CT.

Shoulder MRI can be done to look for and rule out soft tissue shoulder injuries including rotator cuff injury.

Brachial Plexus MRI mainly done to look for and rule out brachial plexus injuries or other soft tissue abnormalities that can impact the nerves such as lipoma, neuroma, or tumors. The test can be performed both with the arms up and with the arms down. There are a couple of non-specific findings that can be suggestive of NTOS but not diagnostic. These would be edema of the brachial plexus or loss of fat surrounding the brachial plexus when the arm is elevated. It can also help with identifying cervical ribs and some scalene muscle abnormalities and fibrous bands. A normal brachial plexus MRI **cannot** rule out NTOS.

Nerve Testing

Nerve testing usually consists of electromyography (EMG) and/or Nerve Conduction Study (NCS). EMG involves the use of very fine small needles inserted into muscles to stimulate the muscle while it records the electrical activity response to that stimulation. NCS involves electrodes being placed on the skin to emit low voltage shocks to activate the nerves while it measures the strength and speed of electrical impulses as they move through nerves. Both tests are for the purpose of measuring how well the nerves and muscles are working. For NTOS, this will usually be performed on the arms. They are mainly done to look for and rule out several other conditions that can mimic NTOS such as carpal tunnel, cubital tunnel, cervical radiculopathy, ALS, or certain nerve injuries.

Most patients with NTOS will have a normal EMG/NCS. The reasons for this are that:

- the nerve compression with NTOS is intermittent and transient which is difficult to capture during a test done over a short period of time.
- unless the nerves are showing permanent damage from the compression, it's unlikely to show up on the EMG and most people with NTOS do not have permanent nerve damage to that degree.
- NTOS involves nerve compression at the level of the brachial plexus which has a much deeper location within the body which also makes it more difficult to test.

Therefore, nerve testing that comes back normal **CANNOT** rule out NTOS. There are situations where EMG/NCS is abnormal in a patient with NTOS, such as:

- When a patient has had longstanding severe compression of the nerves which has resulted in permanent nerve damage. This is pretty rare.
- If a patient has **BOTH** NTOS and another condition such as cervical radiculopathy or carpal tunnel. It is possible to have both conditions. When this occurs, it is usually referred to as "double crush" which is when a nerve is compressed at 2 different places along the same nerve path. For example, if the median nerve is being compressed both at the cervical spine and within the thoracic outlet. For another example, if the median nerve is being compressed both at the thoracic outlet and at the carpal tunnel area close to the wrist. In a double crush scenario, the theory is that when a nerve is compressed at one level, the rest of the nerve downstream from that area sustains swelling, irritation, and is more susceptible to compression injury. Therefore, if a patient has a nerve study which is positive for carpal tunnel or cubital tunnel, but they are exhibiting symptoms which could not possibly be caused by those conditions, then NTOS diagnosis should not be ruled out and should still be pursued.
- If NTOS is compressing a nerve within the thoracic outlet which then manifests itself on nerve testing as carpal or cubital tunnel. Even if not common, sometimes a nerve will show abnormalities on nerve testing that are consistent with carpal or cubital tunnel which are actually being caused by NTOS.
- If the EMG shows abnormality of the Median Antebrachial Cutaneous Nerve (MACN). This nerve has recently been shown to have abnormal findings that relate specifically to brachial plexus compression within the thoracic outlet. The problem is that most doctors performing the EMG are not aware of this and are not trained in how to properly test this nerve. It's typically not part of the standardized testing in most clinics.

Nerve testing abnormalities indicating NTOS are **not** required for diagnosis.

Vascular Testing

Venous or Arterial Doppler Ultrasound During the workup for NTOS, these can be done to help rule out any clots or occlusions if the patient is having symptoms that strongly suggest primary vascular obstructions. Because they are often performed with the arm in several positions, they can also identify if there is loss of blood flow or pulse in these positions. These tests generally are not useful for the diagnosis of NTOS. Whether loss of blood flow or pulse occurs with arms in any elevated position is not clinically significant to NTOS diagnosis. NTOS can occur in patients who do not lose blood flow or pulse with the arms elevated. Also, healthy asymptomatic individuals can lose blood flow or pulse with the arms

elevated. It should also be noted that these studies assess positional arterial compression which is not diagnostic of ATOS and does not explain the resting symptoms which are more likely being caused by NTOS. Therefore, findings of loss of blood flow or pulse with arms elevated should not rule out NTOS in favor of an ATOS diagnosis particularly because NTOS is the more likely diagnosis.

Photoplethysmography (PPG) This test involves infrared sensors being placed on the fingertips which can measure blood flow while the arms are in various positions. It is sometimes done at the same time as doppler ultrasound but can also be done as a standalone test. The same diagnostic limitations and qualifications apply to this test as for the doppler ultrasound as stated above.

CT Angiogram of the Chest this is a CT scan of the chest which can show the vessels particularly the axillary-subclavian vein and axillary-subclavian artery. It is typically done both with the arms up and the arms down. It can help determine location of compression of the vessels, and the condition of the vessels as far as any clots or damage. It is not typically done if a patient is only being worked up for NTOS. However, some TOS specialists do use it for NTOS diagnostic workup, but they only take into consideration compression when the arm is in the down position (at rest). Their interpretation is that, if there is mild compression of the axillary-subclavian artery with the arm at rest, then this can be consistent with NTOS. The reason for this is that the artery and the nerves are side by side as they travel through the thoracic outlet thus if the artery is compressed, then the nerves must be as well. Unless the patient is seeing a TOS specialist who specifically uses this test as part of his or her NTOS workup, it is not a test that would typically be suggested to be done.

PHYSICAL EXAM

A thorough and targeted physical exam is an extremely important aspect of any NTOS diagnostic workup. TOS specialists typically rely heavily on this. There are actually very specific findings on physical exam that are unique to NTOS. Below is a list of what is typically assessed during a targeted NTOS physical exam:

- Resting posture of the patient. NTOS patients will often have posture where the shoulders are rounded and the head and neck are slumped forward.
- Range of motion of the arm as well as whether any symptoms are triggered by these movements.
- Range of motion of the neck – turning side to side and bending ear to shoulder and whether any symptoms are triggered. NTOS symptoms are often triggered by turning the head toward the shoulder of the affected side and by side bending the head ear to shoulder of the non-affected side.
- Hand grip and strength
- Hand muscle atrophy
- Exam of nerves in the lower arm to check for signs of carpal and cubital tunnel. However, it should be noted that if the explanation for symptoms or physical exam findings is that the patient has multiple nerve compression syndromes of the lower arm such as carpal tunnel and cubital tunnel, then it should be considered that the compression is occurring up at the level of the brachial plexus.
- Swelling, temperature or color changes of the arm and hand
- Tenderness of the back of the neck over the cervical spine (may indicate cervical spine disease).
- Increase in symptoms when tilting the head back or when bending the head down toward the chest (may indicate cervical spine disease).
- Tenderness and pain when pushing on the shoulder joint (may indicate shoulder soft tissue disease)

- Shoulder blade winging which can be seen with NTOS but also can be caused by a separate primary nerve injury or entrapment issue.
- Tenderness or deformity of the collarbone joints
- Tenderness of the SCM, upper trapezius, and other upper back muscles including muscles surrounding the shoulder blade
- Tenderness with pushing within the scalene triangle and at the base of the neck just above the collarbone (supraclavicular area) along with whether arm symptoms are triggered by this. This part of the exam can also identify scalene muscle spasm. These findings are unique to NTOS, and this is a very important part of the exam as almost all NTOS patients will have these findings which strongly support NTOS diagnosis.
- Identification of any signs of NPMS, ATOS or VTOS.

PROVOCATIVE MANEUVER TESTING

Provocative maneuver testing is tests involving different arm and/or head & neck movements (maneuvers) which are intended to trigger (provoke) certain symptoms. The two most common tests which are specific to NTOS diagnosis are the Elevated Arm Stress Test (EAST) and the Upper Limb Tension Test (ULTT). Some physicians, typically those who do not specialize in TOS, will use other provocative maneuver tests which are designed to assess positional artery compression where loss of pulse is triggered by certain elevated arm movements. These tests are the Adson's Test and the Wright's Test which are considered to be positive if there is loss of pulse. Loss of pulse does not directly relate to NTOS nerve compression, and these tests also have a high positive rate in normal asymptomatic individuals. In addition, most patients with NTOS will not lose their pulse with arm elevation. Therefore, the Adson's and Wright's tests, when negative, **cannot** rule out NTOS and, when positive, are nonspecific to NTOS.

Elevated Arm Stress Test (EAST) – aka Roos Test Some TOS specialists consider this test to be the most useful provocative maneuver test for NTOS. It involves having the patient raise both arms in a “surrender” position while they repeatedly open and close their hands for 3 minutes. Most patients with NTOS begin to experience reproduction of their symptoms quite quickly within the first minute and are unable to continue to complete the test. If the patient has no symptoms triggered and does not have any difficulty completing the test, it can be interpreted as such that perhaps an alternative diagnosis should be considered.

Upper Limb Tension Test (ULTT) This test involves the patient holding both arms out to their sides at shoulder height with palms facing down. The patient extends both wrists up so fingers are pointing at the ceiling and the neck is side bent toward the non-affected side. Majority of NTOS patients will have symptoms reproduced during some stage of this test.

MUSCLE BLOCK INJECTIONS

It's important to note that a muscle block and a nerve block are not the same thing. Generally, any type of block injection involves using some type of numbing agent such as lidocaine. A nerve block involves injecting lidocaine around a nerve or nerves basically to completely deaden the nerves so that they cannot transmit any signals. With respect to a brachial plexus nerve block, injecting lidocaine around the brachial plexus will only deaden all the nerves which may provide pain relief along with possibly an entirely numb arm, but it will not provide any NTOS diagnostic information. Simply numbing all the brachial plexus nerves will not provide any information about what is causing the symptoms. It just might provide temporary relief. A muscle block involves injecting lidocaine directly into a muscle which deadens the intramuscular nerves. The purpose of this is to temporarily interrupt muscle spasm.

Scalene Muscle Block Because the scalene muscles are often one of the main contributors to NTOS nerve compression which often involves muscle tightness and spasm, a good way to test for NTOS is to stop that spasm and see if there is any symptom relief. Stopping the muscle spasm essentially decompresses the nerves. This is one of the only diagnostic tests that is specific to diagnosing NTOS as opposed to ruling out other medical conditions. Even though both the anterior and middle scalene muscles can cause nerve compression, typically the muscle block is only performed in the anterior scalene muscle. To ensure precise injection location and for safety purposes, it is performed with ultrasound or other imaging guidance such as MRI. The lidocaine is injected directly into the anterior scalene muscle and is typically fast acting within minutes. Most TOS specialists consider a positive response to the block to be obtaining at least 50% relief in symptoms. This measurement does not only apply to pain but to any NTOS symptoms such as range of motion, feeling of arm weakness or fatigue, numbness, tingling, discoloration, or cold sensations, etc. To help the patient with measuring the block response, most clinics will have the patient fill out a pain/symptom diary for a certain number of hours following the block. Sometimes they will have the patient do some provocative maneuvers both before the block and after the block to see if there is any difference in ability or symptoms that are triggered. While the block is active, some patients will try to do certain activities that are usually difficult for them or that trigger symptoms to see if they are better able to do them. Typically, the block only lasts for several hours at most. A positive block response is a very strong indication that NTOS is the proper diagnosis, and some specialists consider it a high likelihood that the patient will respond to treatment whether it is conservative or surgical.

However, a positive scalene block response is **not** required to make a diagnosis of NTOS. In addition, a negative scalene muscle block response **cannot** rule out NTOS and does not mean that a patient will not respond to either conservative or surgical treatment. Some reasons why a patient with NTOS might not get relief from an anterior scalene muscle block are as follows:

- The amount and location of the lidocaine injected might not have been accurate resulting in no spasm interruption or only partial spasm interruption.
- the anterior scalene muscle spasm is not the main cause of the patient's TOS symptoms. Some people with TOS have the middle scalene as the main cause of their symptoms as opposed to the anterior scalene.
- The patient has extensive scarring in the anterior scalene that makes it less responsive to the lidocaine. The lidocaine stops muscle spasm immediately by causing a complete blockade of the intramuscular nerves within the muscle tissues. If the muscle fibers are very scarred which can be common for many with NTOS, then that tissue might not fully respond to the lidocaine. In this case, the nerves within the muscle will not get completely blocked so the spasm never really gets stopped.
- The patient has advanced nerve damage such that even with interruption of the anterior scalene muscle spasm, the nerve symptoms don't get relieved.

Botox Injections Botox injections to the scalene muscles for NTOS are more typically used as a symptom relief measure than as a diagnostic tool. However, some specialists do use them to assist with diagnosis. They are used under the same principle as the lidocaine muscle blocks in that Botox interrupts the muscle spasm and therefore decompresses the nerves resulting in symptom relief. Botox does not take effect as quickly as lidocaine because it works in a different manner, but if effective, it can last up to 3 months. There can be different scenarios under which someone might not respond well to the Botox. Therefore, the same qualifiers apply here as for the lidocaine muscle block, which is that a negative

Botox response cannot rule out NTOS, and it does not mean that a patient will not respond to either conservative or surgical treatment.

PUBLISHED DIAGNOSTIC CRITERIA

In 2016, the top TOS specialists in the United States collaborated to come up with standardized diagnostic criteria for all 3 types of TOS. It was published in an article in the Journal of the Society for Vascular Surgery. Below are the published standardized criteria for the diagnosis of NTOS.

NEUROGENIC TOS

Definition. NTOS should be defined by the presence of *three of the following four criteria*.

1. LOCAL FINDINGS

- a. History: Symptoms consistent with irritation or inflammation at the site of compression—scalene triangle in the case of NTOS and pectoralis insertion site in the case of NPMS—along with symptoms due to referred pain in the areas near the thoracic outlet. Patients may complain of pain in the chest wall, axilla, upper back, shoulder, trapezius region, neck, or head (including headache).
- b. Examination: Pain on palpation of the affected area as above

2. PERIPHERAL FINDINGS

- a. History: Arm or hand symptoms consistent with central nerve compression. Such symptoms can include numbness, pain, paresthesias, vasomotor changes, and weakness (with muscle wasting in extreme cases).
 - i. These peripheral symptoms are often exacerbated by maneuvers that either narrow the thoracic outlet (lifting the arms overhead) or stretch the brachial plexus (dangling; often driving or walking/running).
 - b. Examination: Palpation of the affected area (scalene triangle or pectoralis minor insertion site) often reproduces the peripheral symptoms.
 - i. Peripheral symptoms are often produced or worsened by provocative maneuvers that are believed to narrow the scalene triangle (EAST) or to stretch the brachial plexus (ULTT) (both described later).
3. ABSENCE of other reasonably likely diagnoses (cervical disk disease, shoulder disease, carpal tunnel syndrome, chronic regional pain syndrome, brachial neuritis) that might explain the majority of symptoms
4. In those who undergo it, the response to a properly performed TEST INJECTION is positive.^{8,9}

Illig KA, Donahue D, Duncan A, Freischlag J, Gelabert H, Johansen K, Jordan S, Sanders R, Thompson R. Reporting standards of the Society for Vascular Surgery for thoracic outlet syndrome. *J Vasc Surg.* 2016 Sep;64(3):e23-35.

TREATMENT

General

Treatment of NTOS can be quite difficult given that any type of nerve pain is difficult to treat. But even other symptoms such as muscle pain, tightness, and spasm can be difficult to manage given the fact that there are a lot of different components impacting these symptoms and though many of them are intertwined, there are usually several different areas of the body being impacted all at once. As a whole, types of treatment can vary based on many different factors such as cause of compression, severity of compression, severity of symptoms, types of symptoms, age, impact on quality of life, access to medical care, coexisting medical conditions, and patient preference. The success of any type of treatment can also vary based on those same factors but can also vary based on the fact that each body is different and each situation is unique. Conservative treatment is any treatment that does not involve surgery. The two main types of treatment categories are conservative and surgical.

CONSERVATIVE TREATMENT (NON-SURGICAL)

Activity Modification

Many symptoms of NTOS are triggered by certain movements or activities especially in the early stages of the condition. Therefore, when possible, one of the first steps taken by many patients to control and manage symptoms is to avoid certain aggravating or triggering activities or to modify the way in which they perform the activities. Examples of activities that are known to aggravate NTOS symptoms are overhead activities, lifting a lot of weight, or repetitive arm and hand movements such as typing, sewing, or vacuuming. Therefore, avoiding these activities can provide a lot of symptom relief to some patients. This isn't a great option for someone whose job requires performing these types of activities, so stopping these activities likely isn't a permanent solution for someone in that situation. However, sometimes having temporary restrictions on activities while engaging in other treatment options can manage symptoms enough to eventually be able to go back to those activities later, but this is not the case for everyone. But, in certain situations, for someone whose symptoms are triggered by a hobby or by something that is not vital to their existence, sometimes stopping those activities is all that is needed. No matter what treatment options are being explored, activity restrictions and modifications are usually a part of most NTOS patients' treatment plans. Often, patients can impose helpful restrictions on themselves, but doctors will also provide very specific restrictions for a patient depending on their circumstances. Keep in mind that not all NTOS symptoms are able to be managed by activity modifications.

Medications

Medications are mainly given for symptom control and relief as there are no medications that can remove the physical compression of the nerves. With NTOS, medication treatment often involves a lot of trial and error. There are different types of symptoms in different types of structures which often require multiple medications to treat these areas. The medications used for NTOS are tolerated and handled differently by each person's body. What might work for one person doesn't necessarily work for another. Side effects can be problematic, and a certain medication from a drug class might be more tolerable to someone than other medications within the same class. Therefore, trying several different medications within the same drug class is common. Dosages often must be increased over a period of time or adjusted, so it can take awhile to find the right combination of medications and dosages to suit a particular patient. The trial-and-error process should always be done at the direction of a physician, and dosage adjustments or stopping a medication altogether should never be done without first consulting a physician. Below are the most common types of medication used to treat NTOS:

- **Non-Steroidal Anti-Inflammatory Drugs (NSAIDs)** such as Ibuprofen or Naproxen
- **Muscle Relaxers** such as Robaxin, Flexeril, Baclofen, or Tizanidine
- **Steroids** such as Prednisone (short-term use for flares)
- **Tricyclic Antidepressants** (for nerve pain) such as Amitriptyline, Nortriptyline, or Desipramine
- **SNRI Antidepressants** (for nerve pain) such as Cymbalta or Effexor
- **Anti-Seizure Meds** (for nerve pain) such as Gabapentin or Lyrica
- **Opioid Pain Meds** such as Tramadol, Oxycodone, or Percocet
- **Lidocaine** cream, ointment or patches
- **Compounded Topical Creams/Ointments** – made by a compounding pharmacy where they take several different types of medications and mix them together into a topical cream or ointment
- **Low Dose Naltrexone**

Physical Therapy (PT)

PT is one of the mainstays of NTOS treatment. It is often the first course of treatment right after initial diagnosis. PT for NTOS is much different than PT for other neck/shoulder/arm conditions. Therefore, PT should be with a therapist who has experience with or knowledge of NTOS. This is often very difficult to find as most therapists do not have NTOS experience. However, if the wrong kind of PT is done for NTOS, it can increase symptoms and even cause additional damage to the nerves. For this reason, initial PT is usually attempted for 4-8 weeks with physician follow up and assessment after that time period. If symptoms are worsening and causing significant issues before the end of that time period, it is best to consult with a physician right away. PT for NTOS is usually aimed at the following:

- Scalene muscle relaxation and gentle stretching
- Neck and upper back muscle rebalancing
- Shoulder blade mobility and mechanics
- Diaphragmatic breathing
- Pec minor muscle relaxation and gentle stretching (for those with coexisting NPMS)
- Posture improvement
- Tools to assist with symptoms during activities of daily living i.e. driving, computer work, sleeping

Most TOS specialists have a PT protocol that can be given to the patient's PT if the PT is not very familiar with NTOS. PT will not necessarily work for everyone with NTOS. It is not uncommon for even the right type of PT to worsen symptoms in NTOS patients including to the point of being intolerable especially in those with longstanding compression. Typically, whether PT will help depends on the main source of the patient's nerve compression. If it's mainly postural or occupational or in the early stages of the condition, then there is a better chance of managing symptoms conservatively. However, many patients with NTOS have anatomical anomalies contributing to compression in addition to scar tissue wrapped around the nerves from years of compression, and PT cannot fix those issues. Sometimes, a patient will start out being able to manage symptoms conservatively with PT in addition to other treatments, but NTOS is known to progress if the main underlying cause is not addressed, so then it can become too difficult to manage conservatively. The success of PT is heavily based on the patient's view of the situation and whether they feel that the level of relief obtained in comparison to the level of time and effort required to manage the condition is worth it. Each patient's situation is unique, including which type of therapy helps to manage their condition. Conventional PT is what is most commonly used for treating NTOS. However, some patients find relief in unconventional methods such as Active Release Technique (ART), Feldenkrais, or aquatic therapy.

Botox Injections

Botox is made from botulinum toxin which is the toxin that causes the disease botulism. Used in very small doses, it cannot cause botulism. Most people are familiar with Botox as used for cosmetic purposes to reduce the look of lines and wrinkles on the face. Botox works for that purpose because it temporarily paralyzes muscles into which it is injected. In addition to its cosmetic use, it is also used to treat certain medical conditions. The way it works for treating NTOS is that Botox injected into the scalene muscles interrupts the muscle spasm and therefore decompresses the nerves resulting in symptom relief. It is not a permanent resolution for NTOS because Botox only stays active for so long, and can only be injected every 3 months. Botox injections are usually done by a pain management doctor, interventional radiologist, or physiatrist under ultrasound or other imaging guidance. Following injection, it can take 2-4 weeks for the Botox to kick in and for relief to be felt. Not everyone responds to Botox, so there are some patients who do not get any relief from

it. The time range for which the Botox remains active and effective is different for each patient. There are patients who get significant relief only for a few weeks and there are some who get significant relief for 2-3 months and, therefore, get injections repeated every 3 months. Some specialists believe that, although Botox itself is not a permanent fix for NTOS, PT can more easily be done during the time that the Botox is active and thus longer-term relief can be achieved. There are also patients whose bodies can develop an antibody to the Botox the more often injections are done and thus, over time, the Botox becomes less and less effective. In addition to the scalene muscles, some patients will have other muscles injected that are often impacted by NTOS such as the SCM, upper trapezius or levator scapulae. Botox is not FDA approved to treat NTOS, so most insurance will not cover the cost of it. It can be quite expensive, so checking with the ordering provider and the patient's health insurance company is well advised. There is also a Botox savings program that will reimburse patients who qualify.

Other Miscellaneous Treatment Options

Most NTOS patients are desperate for any kind of pain and symptom relief, and because there can be so many symptoms in different parts of the body, it can often require several different types of treatment to keep things at bay. Once a patient finds a few things that work, they usually only provide very temporary relief and thus must be performed weekly or sometimes even several times a week. Below is a list of some of the different options that can provide symptom relief:

- Trigger Point Injections (lidocaine or steroid injections into muscle trigger points)
- Dry Needling (insertion of needles into muscle trigger points w/o injecting medicine)
- Chiropractor (there are several different types of chiropractic methods. Extreme caution should be used when the diversified method (i.e. cracking/popping) is used on the neck and near the brachial plexus as injury can occur)
- Massage Therapy
- Osteopathic Therapy
- Acupuncture
- Nerve Block Injections
- Cupping
- Topicals such as lidocaine, Biofreeze, IcyHot, Capsaicin, Tiger Balm
- TENS Unit (electrical nerve stimulation via skin electrodes) – should be used with caution and at the direction of a physician or PT as can irritate nerves & cause damage
- CBD products including topicals
- Medical Marijuana (where legal)

It is best to consult with a physician prior to engaging in any forms of treatment.

SURGICAL TREATMENT

Surgery for NTOS is usually offered after all conservative treatment measures have been exhausted to prevent permanent nerve damage and to improve or preserve the patient's quality of life. The surgery is extremely complicated and takes place in a very complex area of the body where several vital structures are all packed tightly together in a very small space. With a surgeon who is **not** experienced in TOS surgery, damage to vital vessels and to nerves that serve the arm and hand can occur and can be devastating. Therefore, of the surgeons available to a patient, it is recommended to choose the surgeon who has the highest level of TOS experience possible. There are very few dedicated TOS centers in the United States and even fewer throughout the rest of the world. Because of this, many patients must travel to other states or other countries for experienced care. However, choosing a highly experienced TOS surgeon gives the patient the highest chance for a

successful outcome and the lowest chance for serious complications. Under these circumstances, TOS surgery can be very safe and successful. For help finding a TOS surgeon, go to www.tosoutreach.com/find-a-surgeon.

What does surgery for NTOS entail?

In general terms, NTOS surgery is often referred to as thoracic outlet decompression surgery as it involves removing anatomical structures and scar tissue in order to decompress the nerves. Surgery typically involves a hospital stay of 2-4 nights on average. There are actually several components to NTOS surgery. Depending on the patient's findings and the surgeon's protocol and experience level, NTOS surgery includes some or all of the following:

- Removal of the first rib known as First Rib Resection (complete or partial)
- Removal of cervical rib, if present, known as Cervical Rib Resection (complete or partial)
- Removal of the anterior scalene muscle known as Anterior Scalenectomy (complete or partial)
- Removal of the middle scalene muscle known as Middle Scalenectomy (complete or partial)
- Removal of scar tissue from around the nerves known as Brachial Plexus Neurolysis (complete or partial)
- Removal of scar tissue from around the axillary-subclavian artery known as Arteriolytic (complete or partial)
- Removal of any fibrous bands which are causing compression
- Removal of any extra scalene muscle or other compressing structures

Most TOS experts believe that performing all of the above is the best way to achieve a complete and thorough decompression and to prevent recurrence of NTOS. However, there are surgeons out there who believe that first rib resection is not necessary or that scalenectomy is not necessary. Because TOS is not a well-known condition and surgery for it is not regularly performed by most surgeons, there exists a wide variety of surgical component combinations that can be performed for NTOS. For this reason, let's break each component down a little further:

Removal of the first rib known as First Rib Resection (complete or partial) As far as history goes, removing the first rib is probably the oldest and most common component of TOS decompression surgery. Most TOS experts believe that all or part of the first rib should be removed for most NTOS patients. Complete removal of the entire first rib would help to decompress the nerves and both vessels of the thoracic outlet. However, when the nerves are the only structure that needs decompressing, as is usually the case with NTOS, then typically only the back $\frac{3}{4}$ (approx.) of the first rib needs to be removed. This is often referred to as the posterior portion of the first rib. Essentially, cutting the rib at the point directly in front of where the anterior scalene muscle attaches to it and removing it from that point all the way back to where the rib attaches to the spine should suffice. Most TOS experts agree that removing any less of the first rib can be problematic as complete decompression of the nerves might not be achieved. In addition to not achieving complete decompression, some TOS experts have also experienced that any amount of posterior rib that is left behind can become an attachment point for post-surgical scar tissue. If scar tissue attaches to the remaining rib, it can compress the nerves again and result in a return of symptoms. Therefore, removal of the first rib can be a twofold component of NTOS surgery – (1) decompressing the nerves and (2) preventing recurrence if scar tissue forms.

Removal of cervical rib, if present, known as Cervical Rib Resection (complete or partial) A cervical rib sometimes contributes to NTOS nerve compression. Most TOS experts will

remove the cervical rib in addition to the first rib. It is usually removed in its entirety. However, certain surgical approaches can limit access to the entire cervical rib and thus partial removal might only be possible. As with the first rib, this might not achieve complete decompression, and can also be a risk to become a scar tissue attachment point which can compress the nerves again and result in return of symptoms.

Removal of the anterior scalene muscle known as Anterior Scalenectomy (complete or partial)

Because the anterior scalene is attached to the first rib, in order to remove the first rib, it must, at a minimum, be detached from the first rib. Whether it also gets removed either partially or completely is up to the surgeon. Detaching the anterior scalene from the rib but **not** removing it is called anterior scalenotomy. Almost all TOS experts agree that either some or all of the anterior scalene muscle should be removed for purposes of complete decompression of the nerves. In addition, some surgical approaches limit access to be able to remove the entire anterior scalene muscle. Some experts argue that partial removal of the anterior scalene is enough to provide long-term significant symptom relief. Other experts argue that, despite any initial symptom relief, symptoms can return later down the road as there is a chance for the remaining part of the scalene to reattach to other structures and again cause compression. This has been a consistent finding during reoperations on patients who had either prior partial anterior scalenectomy or anterior scalenotomy.

Removal of the middle scalene muscle known as Middle Scalenectomy (complete or partial)

Because the middle scalene is attached to the first rib, in order to remove the first rib, it must, at a minimum, be detached from the first rib. Whether it also gets removed either partially or completely is up to the surgeon. Detaching the middle scalene from the rib but **not** removing it is called a middle scalenotomy. Some TOS experts believe that it is unnecessary to remove the middle scalene at all and that simply detaching it from the first rib is enough to provide long-term significant relief. In addition, some surgical approaches limit access to be able to remove the entire middle scalene muscle. Other TOS experts believe that either some or all of the middle scalene muscle should be removed for purposes of complete decompression of the nerves particularly since there are a few nerves that run directly through the middle of the middle scalene. Those same experts argue that, as with the anterior scalene, despite any initial symptom relief, symptoms can return later down the road as there is a chance for the remaining part of the scalene to reattach to other structures and again cause compression. This has been a consistent finding during reoperations on patients who had either prior partial middle scalenectomy or middle scalenotomy.

Removal of scar tissue from around the nerves known as Brachial Plexus Neurolysis (complete or partial)

Almost all TOS experts agree that either partial or complete neurolysis of the brachial plexus nerves should be done for complete decompression. Complete neurolysis means that scar tissue has been removed from all five brachial plexus nerve roots. Some surgical approaches limit the access needed to remove scar tissue from all nerve roots which usually means that only the lower three roots have scar tissue removed. Some TOS experts argue that partial neurolysis should be enough because the lower three nerve roots are the nerve roots most commonly affected by NTOS. Other TOS experts argue that all five nerve roots should have scar tissue removed because all nerve roots can be affected by NTOS and that any nerve roots left with scar tissue encasing them would not be fully decompressed and would therefore continue to cause symptoms.

Removal of scar tissue from around the axillary-subclavian artery known as Arteriolytic (complete or partial)

Even though a patient might be primarily undergoing surgery for TOS nerve compression, it is not uncommon for these patients to also have some level of scar tissue around the axillary-subclavian artery as the nerves and the artery are right next to each other. This is not considered to be ATOS, but most experts will go ahead and remove any scar tissue from around the artery if they see it while decompressing the nerves.

Removal of any fibrous bands which are causing compression Fibrous bands are commonly found connected to different structures within the thoracic outlet of many NTOS patients. Most TOS experts would agree that any fibrous band contributing to compression should be removed. However, depending on where a band starts and ends, some surgical approaches might limit access to be able to remove it. As with the scalene muscles, some TOS experts believe that partial removal of a fibrous band comes with the risk for it to reattach to other structures and again compress the nerves resulting in return of symptoms.

Removal of any extra scalene muscle or other compressing structures The most common structure to fall into this category is an extra scalene muscle known as a scalene minimus. Most TOS experts agree that removal of a scalene minimus muscle should be performed for complete decompression. However, depending on where it attaches, there is a small likelihood that it might not be fully accessible with certain surgical approaches.

As with any surgery, the decision to have surgery and which components should be included in said surgery should be made based on advice given by the patient's surgeon as to what is best for their specific clinical situation.

NTOS Surgery Recovery

In general, recovery from NTOS decompression surgery is a bit different from most more common surgeries due in large part to the fact that it involves the brachial plexus nerves. Your run of the mill orthopedic surgery or gallbladder surgery recovery usually involves a linear steady line of progression with the worst being day 1 of surgery with steady improvement from that time forward over the course of several weeks. This is usually not the case with NTOS surgery. Nerves, especially near the brachial plexus, have a very complicated and somewhat turbulent regeneration process. Complete nerve healing for sites at the brachial plexus can take up to 2 years or longer. The recovery trajectory is typically not linear. Instead, it bounces around more like a roller coaster. It is very difficult to predict what each patient's recovery will be like, and it is different for each patient. Here are some factors that can affect a patient's recovery:

- Age. Usually, the younger the patient, the faster and easier the recovery is.
- How long the nerves have been compressed
- How bad the compression and symptoms are
- How much scar tissue there is to be removed
- How sensitive the patient's nervous system is
- What is done during surgery
- Experience and skill of the surgeon

Following NTOS decompression surgery, the nerves will be decompressed, but this does not mean that they are healed. The typical NTOS patient has nerves that have been compressed for several years, and despite being decompressed, they still need to heal from the damage sustained from the compression. The nerves also get worked on and manipulated during surgery particularly when scar tissue is removed from them, so they must heal from this as well. For this reason, many patients will experience pain and symptoms that are temporarily worse than before surgery which is normal. Below is a list of expectations following NTOS surgery keeping in mind that every recovery is different and not everyone will experience the things listed.

Symptoms

- can and will include original symptoms the patient had pre-op in addition to new symptoms caused by the surgery
- some pre-op symptoms can **temporarily** be worse than before surgery
- new symptoms can be felt in the neck, chest, upper back, shoulder, arm, or hand
- new symptoms from surgery can include numbness, tingling, pain, burning, skin hypersensitivity, itching, stinging, electrical zaps, cold sensations, hot sensations, skin color and temperature changes, muscle twitching, neck and upper back muscle tightness, inability to use arm, hand or fingers
- can be unpredictable – symptoms can change daily in type, location, and/or intensity. New symptoms can pop up weeks or even several months into recovery as the nerves enter different phases of regeneration.
- Some loss of arm strength and range of motion. Some can have full range of motion immediately after surgery, and some can have very little to no range of motion. But most fall somewhere in the middle. Most do PT to regain arm strength and range of motion.
- Immediately following surgery, shoulder blade pain is usually the most complained about symptom. Almost every patient has this.
- Low energy and general fatigue with activity. For some, this can last for several months.
- Can take several months to notice significant improvement and for some patients up to a year or longer
- Not uncommon to still experience some level of symptoms including flares of symptoms during the entirety of the healing phase

Pain Patterns

- In general, pain-wise, usually the first 4-6 weeks are the worst
- Most patients generally fall within one of these groups:
 - o Pain is immediately quite bad right after surgery & stays that way for the first several weeks
 - o Pain is not too bad initially but slowly increases peaking around days 7-10 post op & stays that way for first several weeks
 - o Pain is relatively mild until around 3 weeks post op when it spikes & stays that way through week 8
- The difference in pain patterns is usually due to the timing of the nerves “waking up” after surgery. The first step in nerve healing is usually a dormant resting phase prior to the regeneration phase. The symptoms tend to get worse once the regeneration phase begins.
- Usually a cycle of “good” days and “bad” days where a patient can feel like they are really making progress and then the next day feel like they are completely back at square one. Eventually, there are more good days than bad days.

Restrictions

- Most return to work anywhere from 4-12 weeks post op depending on how physically demanding the job is and how well the nerves are healing
- Most have weight restrictions of no more than 5-10 lbs for at least 4-6 weeks. This usually includes lifting, pushing, pulling, etc.
- Most have activity restrictions including no overhead or above shoulder activity and nothing repetitive for at least 4-6 weeks
- Most are able to return to driving by 4 weeks post op

- Restrictions are at the discretion of the surgeon as to when they are imposed and when they are lifted. It can be different for everyone as the surgery and recovery is not a one size fits all.
- Putting too much stress on the nerves and overworking them can lead to delayed healing and risk of scar tissue formation around the nerves leading to compression and resulting in return of symptoms. Therefore, any activity that causes a significant increase in pain and symptoms (particularly nerve symptoms) should either be avoided or done with extreme caution. This includes PT which should be discussed with the patient's surgeon and physical therapist.

Despite the fact that NTOS surgery recovery can be difficult and long, the ultimate outcome is usually more than worth it. Surgery is not a quick fix, and the recovery often requires commitment, discipline and patience, but it can give patients their lives back.

Return of Symptoms (Recurrent TOS)

Sometimes a patient will experience initial relief and then have symptoms return. The two most likely causes for a recurrence of symptoms are:

Complete decompression not achieved during surgery this would occur as described earlier on this page when some anatomy, usually rib or scalene muscle, was either not removed at all or only partially removed.

Scar tissue formation around the nerves this is most likely to occur within the first 1-2 years post op. It can occur from the patient overusing the nerves too much too soon while they are still healing. It can also form if a secondary injury occurs such as a fall or a car accident or any injury to the chest, neck, shoulder, or arm. The injury can reignite the healing process within the surgical field and an overgrowth of scar tissue occurs. It can also form in those patients who have a genetic predisposition to make a lot of scar tissue.

For more information on all types of TOS including symptoms, diagnosis, treatment, and more, please visit www.TOSOutreach.com.