DIAGNOSTIC AND THERAPEUTIC MANAGEMENT OF THE THORACIC OUTLET SYNDROME

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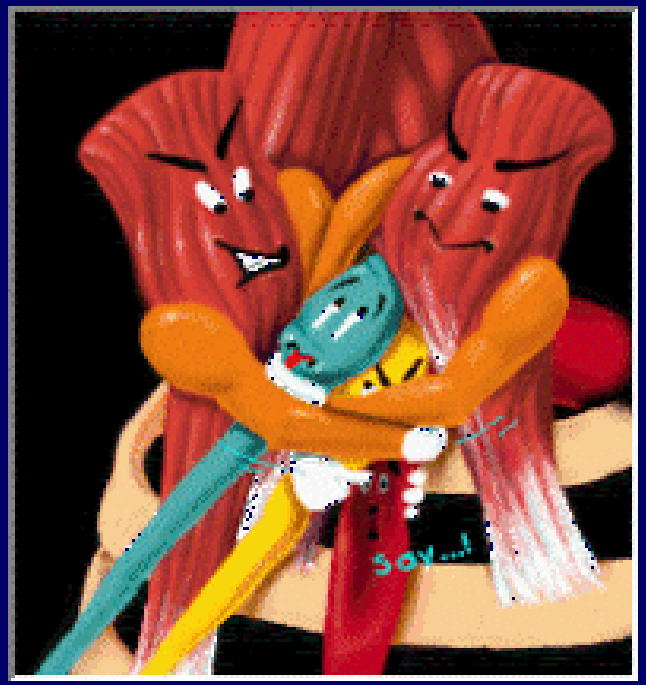
SUPERIOR THORACIC OUTLET

The superior thoracic outlet is the anatomical area crossed by the brachial plexus, and by the subclavian artery and vein. It lies between the anterior and middle scalene muscles, superiorly to the first rib, posteriorly to the clavicle, laterally to the sternal manubrium



Thoracic Outlet Syndrome, TOS

- Incidence 0.3-8%
- Predominantly young people (20-50 y)
- Predominantly female patients (60-70%)
- Predominantly neurogenic (nTOS, 90-95%); 5-10% vascular (1-2% arterial, aTOS; 3-5% venous,vTOS)
- Both aTOS and nTOS share common etiologies causing artery and/or nerve compression, such as trauma (whiplash injury), or anatomic abnormalities (cervical ribs, anterior and/or middle scalene hypertrophy, tumors, or fibrous bands); vTOS is more common in athletes (e.g., volley, baseball, swimming, body-building, etc.), manual workers or subjects performing vigorous activity
- Due to the non-specific nature of signs and symptoms, to the lack of a consensus for the objective diagnosis, and to the wide range of etiologies, the actual figure is still a matter of debate
- Need of a multiple clinical and instrumental diagnostic approach

MULTIDISCIPLINARY TASK FOCE

frontiers in Cardiovascular Medicine

BRIEF RESEARCH REPORT
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Diagnostic and Therapeutic Management of the Thoracic Outlet Syndrome. Review of the Literature and Report of an Italian Experience

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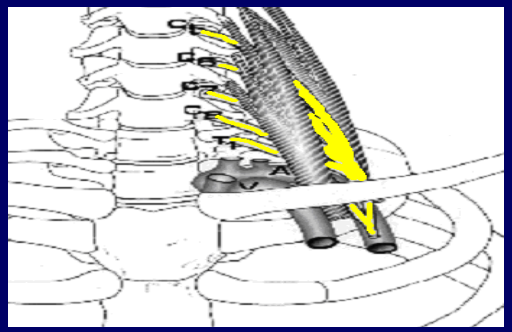
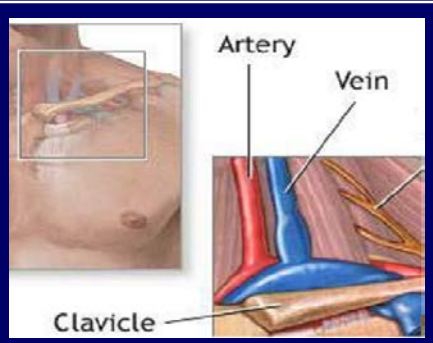
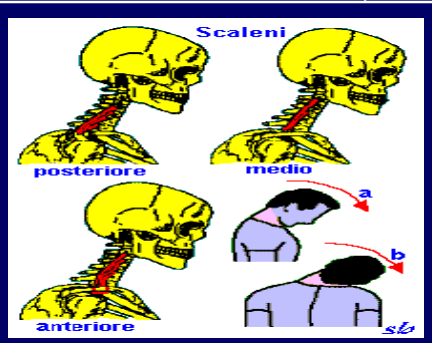
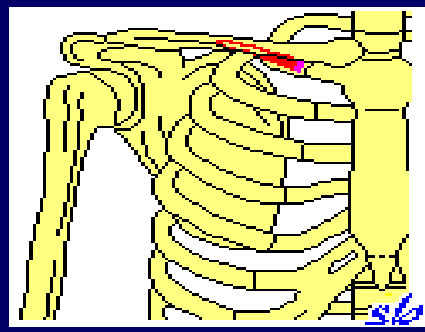
ANATOMY

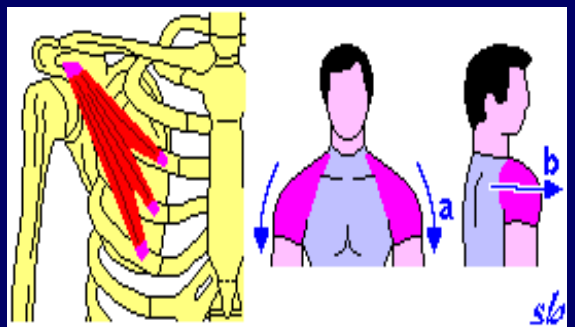
Triangle of the scalenes	Anterior scalen muscle (ant), middle scalene muscle (post), 1st rib (base)	Compression of the subclavian artery and brachial plexus
Costo-clavicular space	Middle third of the clavicle (ant), 1st rib and aponeurosis of the subclavian muscle (post-med)	Compression of the Subclavian vein
Subpectoral space	Tendon of the pectoralis minor muscle and coracoid process (ant), thoracic wall (post)	Compression of the entire brachial plexus

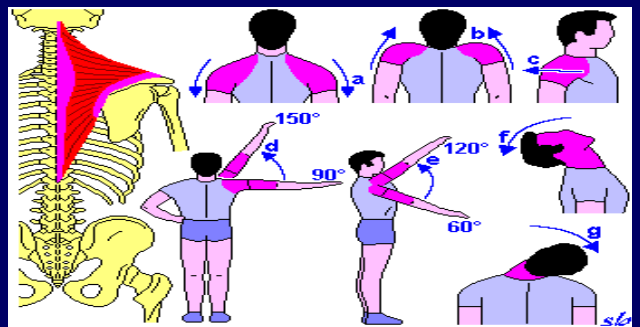












APPROCCIO CLINICO

TABLE 1 | Common causes of TOS.

Congenital factors	Acquired abnormalities
Cervical rib	Postural factors
1st rib anomaly	Fall injuries to upper limb
C7 transverse process abnormalities	Clavicular Fracture
Fibrous bundles between transverse process of C7 and the 1st rib	1st rib fracture
Supernumerary rib	Whiplash injury
Anomalies of scalene muscle insertion	Repetitive stress injuries
Supernumerary scalene muscle	Hypertrophy of the scalene muscles
Exostosis of the first rib	Decrease trapezius, scapulae elevator, rhomboides muscles tone
Cervicodorsal scoliosis	Shortening of the scalene, trapezius, elevator scapulae, pectoralis muscles

Clinical diagnosis

Table 1S. List of signs and symptoms according to the type of TOS

TOS	Anamnesis	Signs and symptoms
nTOS cervical plexus compression	Chronic worsening onset	pain paresthesia
	Repetitive movements	weakness and atrophy of thenar, hypothenar and interosseous muscles eminence, loss of dexterity
	Athletes	occipital headache
	Cervical spine trauma	altered tone of trapezius, scalene, pectoralis major/minor, elevator of the scapula, sternocleidomastoid, serratus anterior muscles Superior plexus (C5-C7): pain in the neck, shoulder, chest and supraclavicular region, arm weakness, paresthesia of the I, II and III finger; Inferior plexus (C7-T1): pain in the medial region of the arm, forearm and hand, paresthesia IV- V finger
vTOS subclavian vein compression	Acute onset Upper limb vein thrombosis Exacerbated by lifting weights above the head.	upper limb edema cyanosis feeling of heaviness in the arm pain temperature alteration
aTOS subclavian artery compression	Cervical rib Acquired in professional weightlifters	Chronic: pain reduction or disappearance of radial pulse following arm movements paleness pulsations, blowing in the supraclavicular area (aneurysm) difference of blood pressure between the limbs weakness and fatigue
		temperature alteration Acute: ischemia of the hand in case of distal embolization

SINTOMATOLOGIA

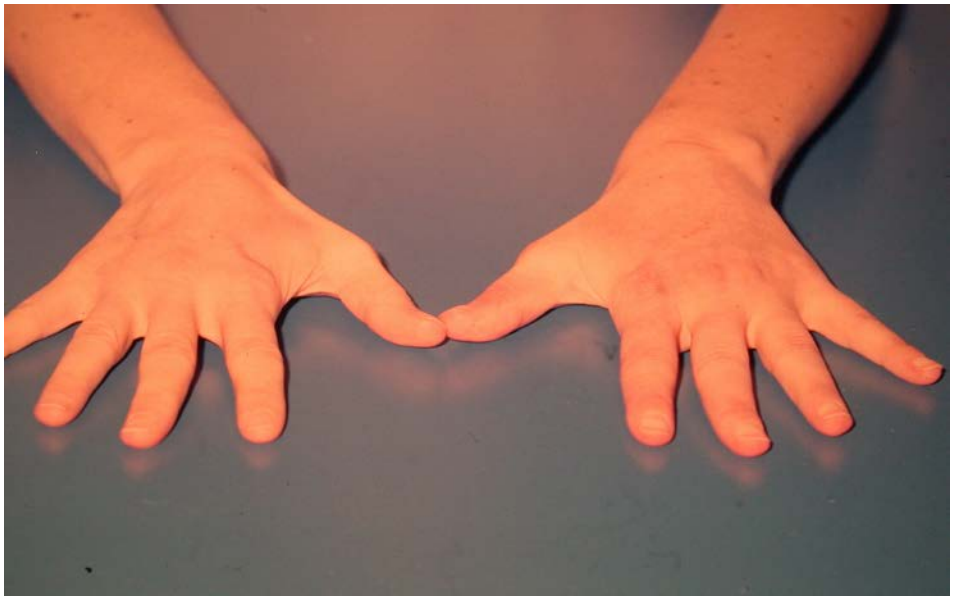
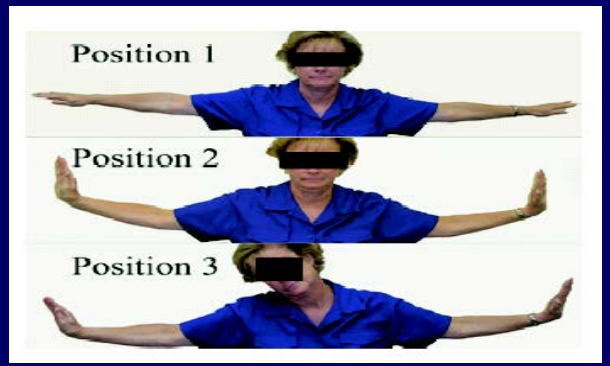


Table 2S. List of provocative maneuvers for clinical diagnosis of TOS

Test	Procedure	Positive Test
Adson Test	The examiner monitors the radial pulse at wrist while the patient's arm is held in extension, the shoulder in external rotation and 30° abduction. Patient takes a deep breath and turns head toward the test arm while extending the neck.	Decrease/disappearance of radial pulse.
Modified Adson Test	The examiner locates the radial pulse, the shoulder is abducted at 90° with extended elbow. Patient takes a deep breath and turn head away from the side tested	Decrease/disappearance of radial pulse.
Upper Limb Neural Tension Test (ULTT) **or modified test of Elvey**	Shoulder girdle depression, shoulder abduction and external rotation, elbow flexion, wrist and Finger extension	Onset of paresthesia
Elevated Arm Stress Test (EAST) **Roos Test**	Patient abducts shoulders to 90 °, externally rotates the shoulders, and flexes the elbows to 90°. Then he opens and closes the hand slowly for three minutes.	Inability to complete the test or experiences heaviness, numbness, tingling or pain
Halstead Maneuver	The examiner palpates the radial pulse and applies downward traction on the test extremity while the patient's neck is hyperextended and rotated to the opposite side.	Decrease/disappearance of radial pulse.



Eden Test	The patient is sitting with the back straight and the shoulders pushed backward and downward. The examiner depresses the shoulder while monitoring the radial pulse at the wrist.	Decrease/disappearance of radial pulse.
Wright Test	The examiner brings the arm in abduction and external rotation to 90° without tilting the head. The elbow is flexed no more than 45°. The arm is then held for 1 min.	Decrease/disappearance of radial pulse. Onset of pare-sthesia

Simultaneous positivity of several maneuvers may increase specificity; for instance, in a study by Gillard et al. the specificity of the Adson and Roos tests ranged from 30 to 72% when used alone, increasing to 82% when both were performed

INTRUMENTAL DIAGNOSIS 1ST LEVEL

- **Finger plethysmography:** may detect a subclavian artery compression displaying both a delayed upslope of the sphygmic wave, and a loss of the dicrotic notch during provocativemaneuvers. These findings, however, are also common in the normal population, mandating further diagnostic testing
- **Plain chest and/or cervical X-ray:** should be obtained in all patients with suspected TOS, being a sensitive and low-cost modality to identify major bone abnormalities potentially causing TOS
- **Electromyography:** investigation of the brachial plexus by measuring the nerve conduction velocity at rest and during activation test: positive if velocity < 60 m/sec

INSTRUMENTAL DIAGNOSIS

2ND LEVEL

Computed Tomography Angiography

CTA is especially useful in symptomatic patients without skeletal anomalies at conventional radiography. Several studies have focused on the efficacy of 3-D reconstructions, which can define the anatomical details, highlight anatomical relationships between vascular and bone structures, and allow surgery planning. CTA is the preferred choice for the evaluation of patients with suspected anomalous ribs or fractures, and it is also useful in postoperative patients with suspected remnant 1st rib.

Among CTA limitations are: ionizing radiation exposure, scarce visualization of brachial plexus, mandatory supine position, and poor arm/shoulder hyperabduction due to the gantry size

INSTRUMENTAL DIAGNOSIS

2ND LEVEL

Magnetic Resonance Angiography

Test of choice if suspected TOS for careful study of all the anatomical components of the thoracic outlet. Even without contrast media, MRA can show arterial and neural plexus compression, venous thrombosis and collaterals, muscle hypo/hyperthrophy, accessories muscles, and anomalous fibrous bands.

- T1-weighted sagittal sequences: vascular and neural compression, post-stenotic dilation;
- Coronal sequences: brachial plexus, fibrous bands.

All sequences on bilateral arm abduction with the head and neck in the neutral position, and repeated during arm adduction with additional contrast administration.

Some advantages over CTA: multi-planar analysis, optimal small parts visualization, lack of ionizing radiation. When not performed due to claustrophobia or implanted devices, CTA remains the study of choice.

Both MRA and CTA are difficult to perform in patients with severe or dialysis-dependent renal failure.

Color-Coded Doppler Ultrasonography

Allows for non-invasive real-time visualization, as well as for pulsed Doppler waveform analysis and blood-flow velocity evaluation, of the subclavian and axillary vessels, both at rest and during provocative maneuvers.

Currently, a consensus on the preferred technique to evaluate suspected TOS by ultrasonography is lacking. In our Institution, all sonographic examinations are performed with last generation ultrasound equipment, using a 3–13 MHz multifrequency linear-array probe for higher accuracy, according to a standardized technique

With the patient supine, DVT of the upper limb veins is ruled-out by CUS. If DVT is excluded, the patient is allowed to sit with the back and neck straight, and with the shoulders resting in a neutral position. The probe is placed longitudinally under the clavicle and the PRF is lowered to a slow-flow velocity program for vein flow detection. Once normal venous flow is detected, provocative maneuvers are started, with the arm being passively abducted at 90°/180°. A nurse standing behind the bed supports the patient's arm during the maneuvers to avoid muscle contraction, potentially altering blood-flow dynamics. The finding of a "paradoxical obstruction" of the deep venous outflow, characterized by a complete flow-stop of and by a dilatation of the subclavian vein proximal to the compression site is diagnostic for significant vein compression. Arterial flow is investigated at the axillary artery, because the subclavian artery runs below the clavicle and, during the provocative maneuvers, it is difficult to insonate it properly. Again, the axillary artery is explored with the arm abducted at 90°/180°. A significant compression of the subclavian artery translates in a damped (monophasic or biphasic), or in a complete Doppler waveform loss recorded in the axillary artery, according to the grade of compression.

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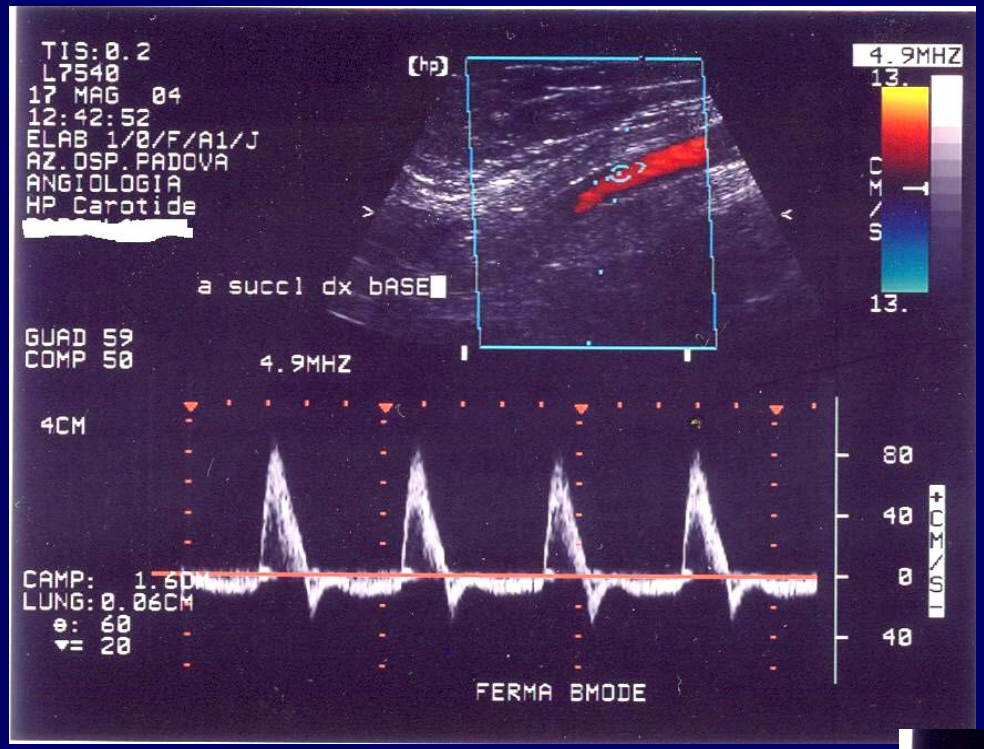
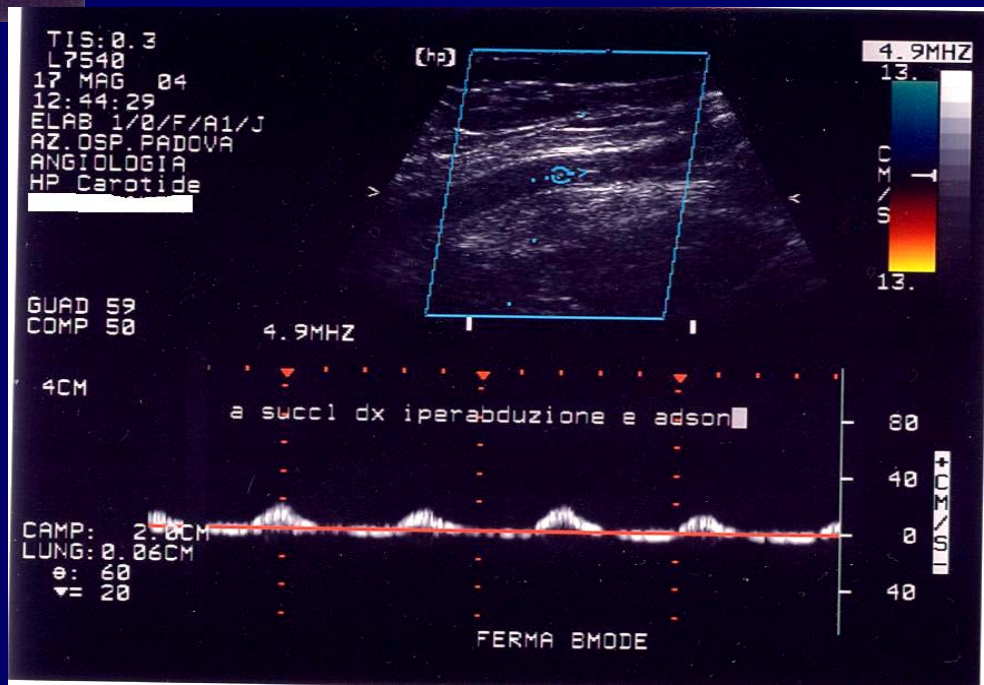


Basal

During provocative maneuvres



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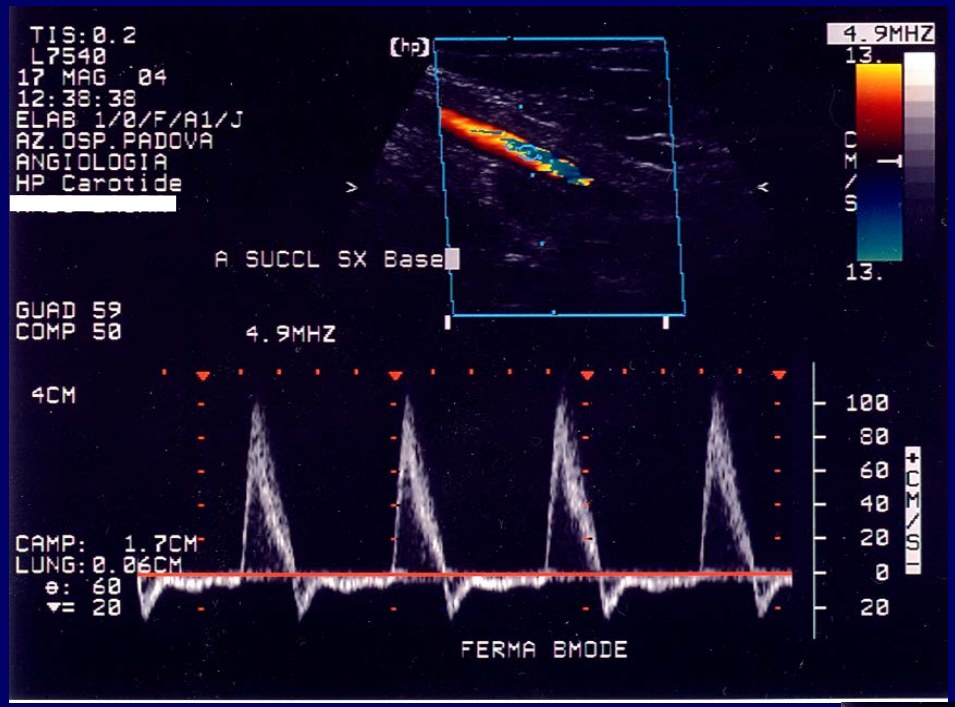

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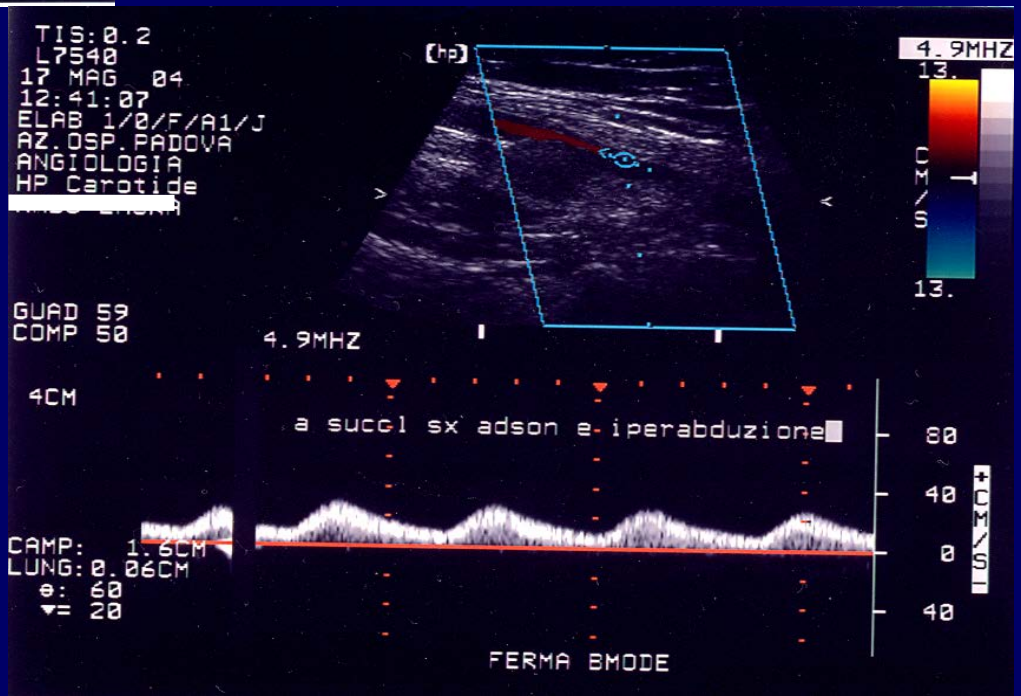
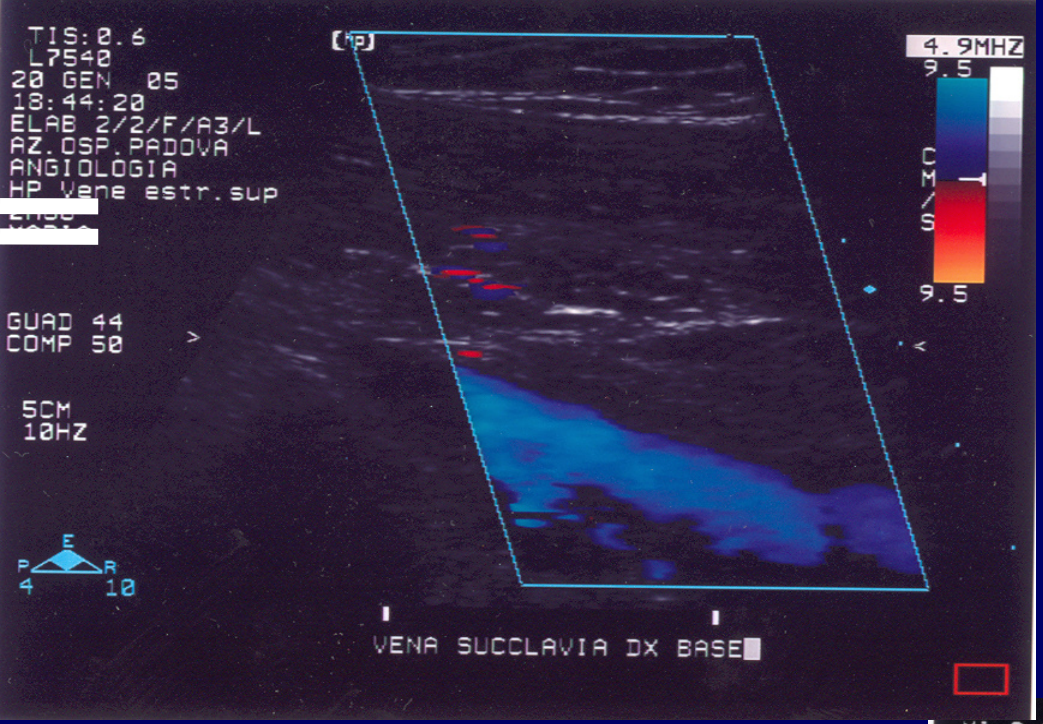
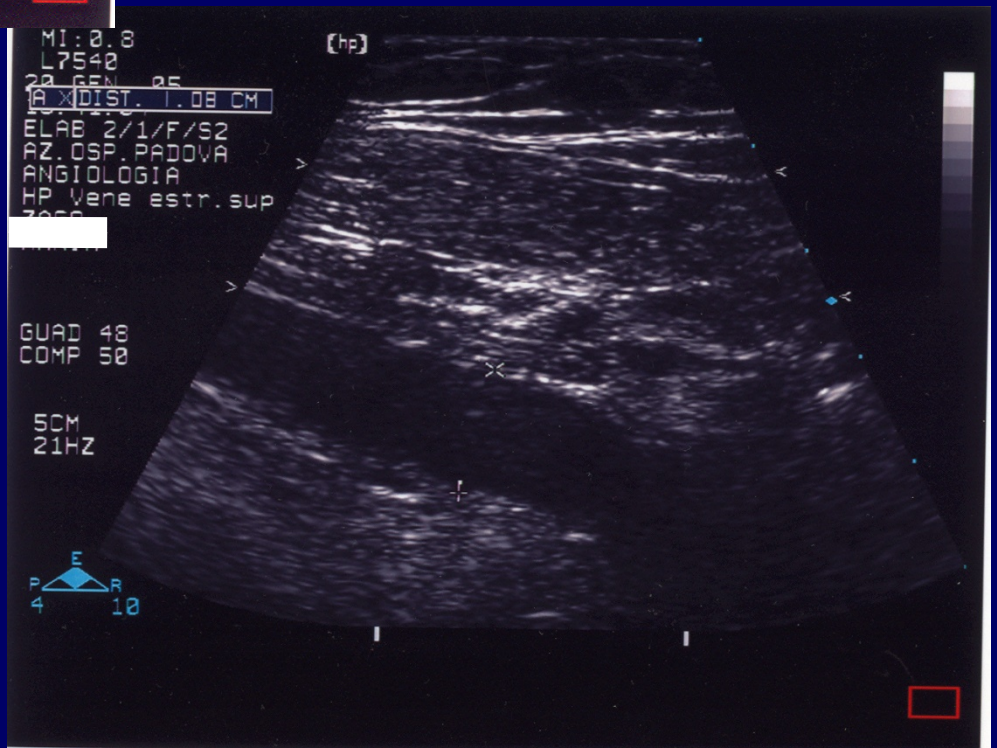
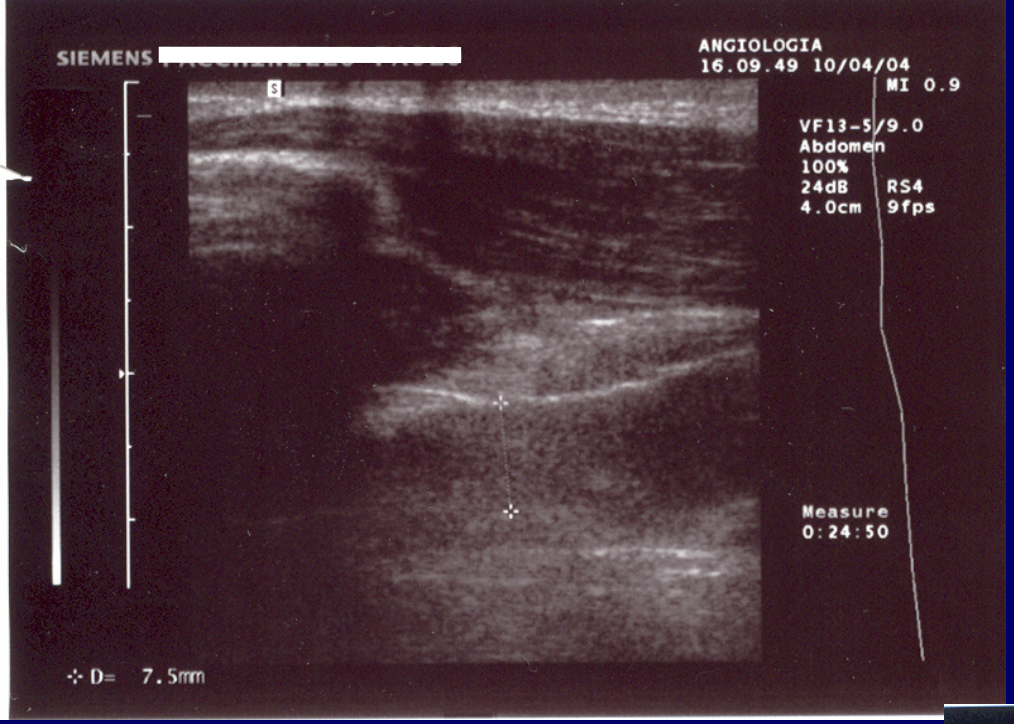




Subclavian vein at rest

Subclavian vein during hyperabduction of the arm at 90°





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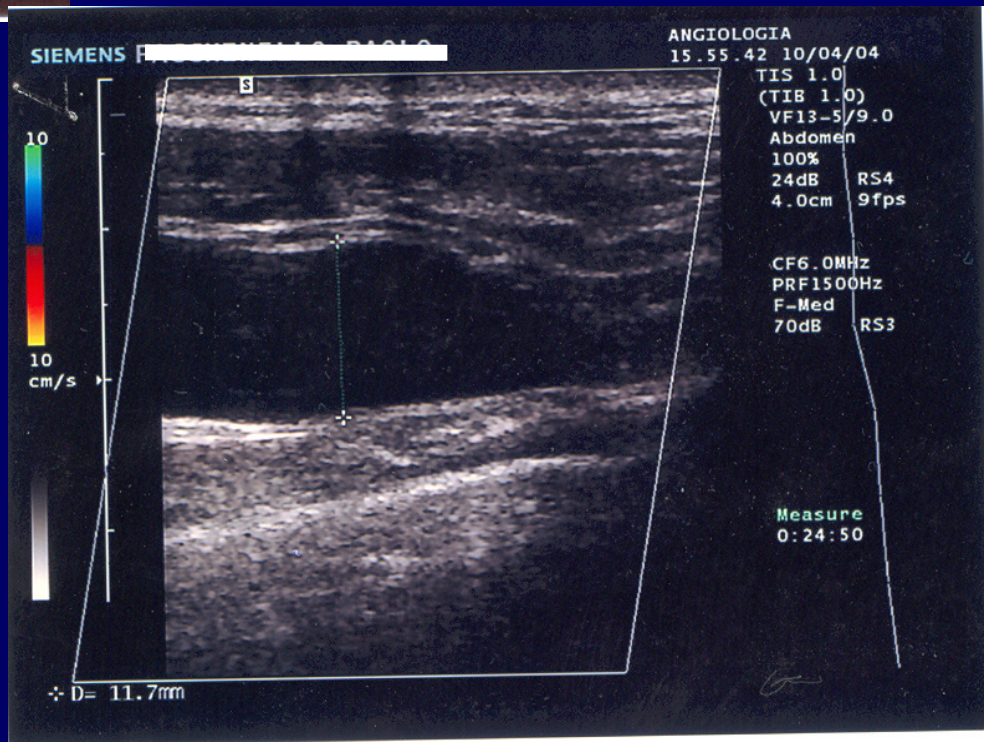

TREATMENT

- Initial management of TOS is usually conservative (dedicated physical therapy, addressing muscle imbalance, postural abnormalities and neural mobilities). Patients are taught that overhead activity, heavy lifting, repetitive motions or use of vibratory tools will aggravate their symptoms, and play against good long-term physical or surgical results.
- Although a consensus on the appropriate conservative regimen for nTOS remains controversial, a multimodal approach including patient education, **TOS-specific rehabilitation and drug therapies has shown positive results in 60–90% of cases.**
- If symptoms persist after at least 3–6 months of rehabilitation and patients are experiencing some degree of disability at work, sleep, recreation, or daily living activities; or in case of worsening symptoms despite rehabilitation, a surgical approach should be considered, and treatment choice is usually related to surgeon experience, kind of involved anatomical district, extent of surgical procedure, and exposure needs.
- Other indications for surgery include arterial and/or venous compression with or without parietal damages, thrombosis or aneurysms

CONSERVATIVE REHABILITATIVE TREATMENT

- Main purpose: to restore the width of anatomical spaces, whose compression is at the basis of the pathology; support the diagnosis of TOS, if symptom improvement is observed. Physical therapy is associated with significant symptom improvement in 60-90% of patients.
- A 6-month program consisting of home-exercises, stretching, postural corrections, and muscle recruitment patterns, primarily focusing on the neck and shoulder, can alleviate TOS symptoms. Generally, patients with mild TOS are expected to improve within 6 weeks.
- In refractory cases undergoing surgery, post-surgical rehabilitation plays a key role in the recovery of autonomy and upper limb range of motion, and in the improvement of the patients' quality of life.

CONSERVATIVE REHABILITATIVE TREATMENT

- Key points of rehabilitative treatment are: postural education (e.g., avoid carrying heavy weights and prolonged hyperabduction of the upper limbs); cervico-dorsal and scapular girdle massage (to resolve contractures); diathermy or laser therapy (for antalgic purposes); kinesiotherapy (to restore the balance between muscles opening and closing thoracic egress).
- The rehabilitation course should be scheduled as follows: postural exercises; static reinforcement of the muscles that open the strait; stretching of the muscles that close the strait; kinesiotherapy of the cervical spine; breathing exercises to lessen the overload of scalene muscles and to lower the first rib. The rehabilitation program must be guided by a physiotherapist specialized in TOS treatment.

SURGICAL TREATMENT

Vascular surgery

- The three main concepts of vascular surgical treatment are: relieving the arterial compression (the trigger of the disease), repairing the damaged subclavian artery (local complication), and restoring the distal circulation (distal complication).

The indications for vascular surgery are:

- failure of conservative therapy with persisting disabling symptoms that interfere with daily life activities;
- vascular (arterial) complications, such as: stenosis, thrombosis, distal embolization or aneurysm

SURGICAL TREATMENT

Thoracic surgery

- In properly selected patients, clinical results of first rib resection may be considered good (complete relief of symptoms) in 85% of patients, fair (improvement with some residual or recurrent mild symptoms) in 10% and poor (no change, or rarely even worsening from preoperative status) in 5%.
- Recently, removal of the first rib on total videothoracoscopic or robotic approach was described, but the outcomes are yet to be completely determined
- Considering the peculiar anatomical district, there are many possible complications and, although rarely, they may also be very serious

NEUROLOGIC COMPLICATIONS

- **Brachial plexus injury**: due to excessive traction to the roots of the plexus during mobilization of first rib; to reduce this risk, it is useful to raise the shoulder and to bend the head toward the operative side.
- **Phrenic nerve injury:** may occur with just minor traction or during a lifting with a forceps, so every contact should be avoided or limited, even with a vessel loop. Another kind of damage is the contact with the cautery, uni- or bipolar.
- **Long thoracic nerve injury:** may occur by cutting one of branches of the nerve, usually running near the lateral side of middle scalene muscle, causing a winged scapula
- **Thoracic duct injury:** the thoracic duct may lie in the middle of the scalene fat pad in the lower left portion of the neck; injury at this level causes milky (or clear) fluid leaking in the operative field. If a leak is evident, damage is managed by ties, clips or bipolar cautery.

VASCULAR COMPLICATIONS

- Raynaud phenomenon with or without ischemic lesions at fingers
- Aneurysm of the subclavian artery (possibly with thrombo-embolic complications -"blue toe syndrome") pre- or post- the compression site
- Aneurysm of the subclavian vein (rare)
- Deep Vein Thrombosis of the upper limb (with superficial compensatory reticular collateral veins at the shoulder and/or at the chest)

TABLE 2 | Demographics of the investigated population.

		Treatment		
		No (39)	**Yes (285)**	***p***
Age	(years, mean + SD)	39.6 ± 11.7	37.9 ± 11.7	0.405
Sex	F	32 (82.1)	238 (83.5)	0.820
	M	7 (17.9)	47 (16.5)	
TOS variant	aTOS	1 (2.6)	12 (4.2)	0.254
	vTOS	1 (2.6)	22 (7.7)	
	vaTOS	4 (10.3)	38 (13.3)	
	nTOS	17 (43.6)	77 (27.0)	
	mTOS	16 (41.0)	136 (47.7)	
Job	High risk workers[a]	9 (23.1)	101 (35.4)	0.151
	Low risk workers[b]	30 (76.9)	184 (64.6)	
Comorbidities	C7 abnormalities	6 (15.3)	42 (14.7)	0.991
	Shoulder disorders	6 (15.3)	39 (13.7)	
	Whiplash	5 (12.8)	36 (12.6)	
	Previous dvt upper limbs	9 (23.1)	68 (23.9)	
Conservative treatment	Massages		20 (7.0)	
	Massages + specific TOS m&ph rehab protocol		74 (26.0)	
	CTEN stimulation		17 (6.0)	
	CTEN stimulation + specific TOS m&ph rehab protocol		60 (21.1)	
	hydrogalvanotherapy		15 (5.3)	
	Hydrogalvanotherapy + specific TOS m&ph rehab protocol		53 (18.6)	
	Specific TOS m&ph rehab protocol		46 (16.1)	
Surgical Treatment	Cervical rib resection		4 (1.4)	
	Cervical rib resection + neurolysis		3 (1.1)	
	Cervical rib resection + scalenectomy		2 (0.7)	
	First rib resection		11 (3.9)	
	First rib resection + neurolysis		2 (0.7)	
	First rib resection + scalenectomy		1 (0.4)	
	Neurolysis		1 (0.4)	
	Other surgery		3 (1.1)	
	Scalenectomy		2 (0.7)	

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Treated and untreated patients were evaluated by the Numeric Pain Rating Scale (NRS) to assess pain burden, either at baseline (T0), after 6 months (T1), and at the last available follow-up visit (T2). Three groups of patients were identified: worsened symptoms (NRS value at T-2 greater than at T-0); stationary symptoms (no difference between T-2 and T-0 NRS values); improved symptoms (NRS value at T-2 lower than at T-0)

TABLE 3 | Symptoms variation in patients undergoing a specific rehabilitation protocol vs. those refusing treatment at the last available follow-up visit.

	Control group (39)	TOS specific rehabilitation protocol (285)	*p* value*
Improved, *n* (%)	21 (53.8%)	192 (67.4%)	<0.01
Stationary, *n* (%)	0	72 (25.3%)	
Worsened, *n* (%)	18 (46.2%)	21 (7.4%)	

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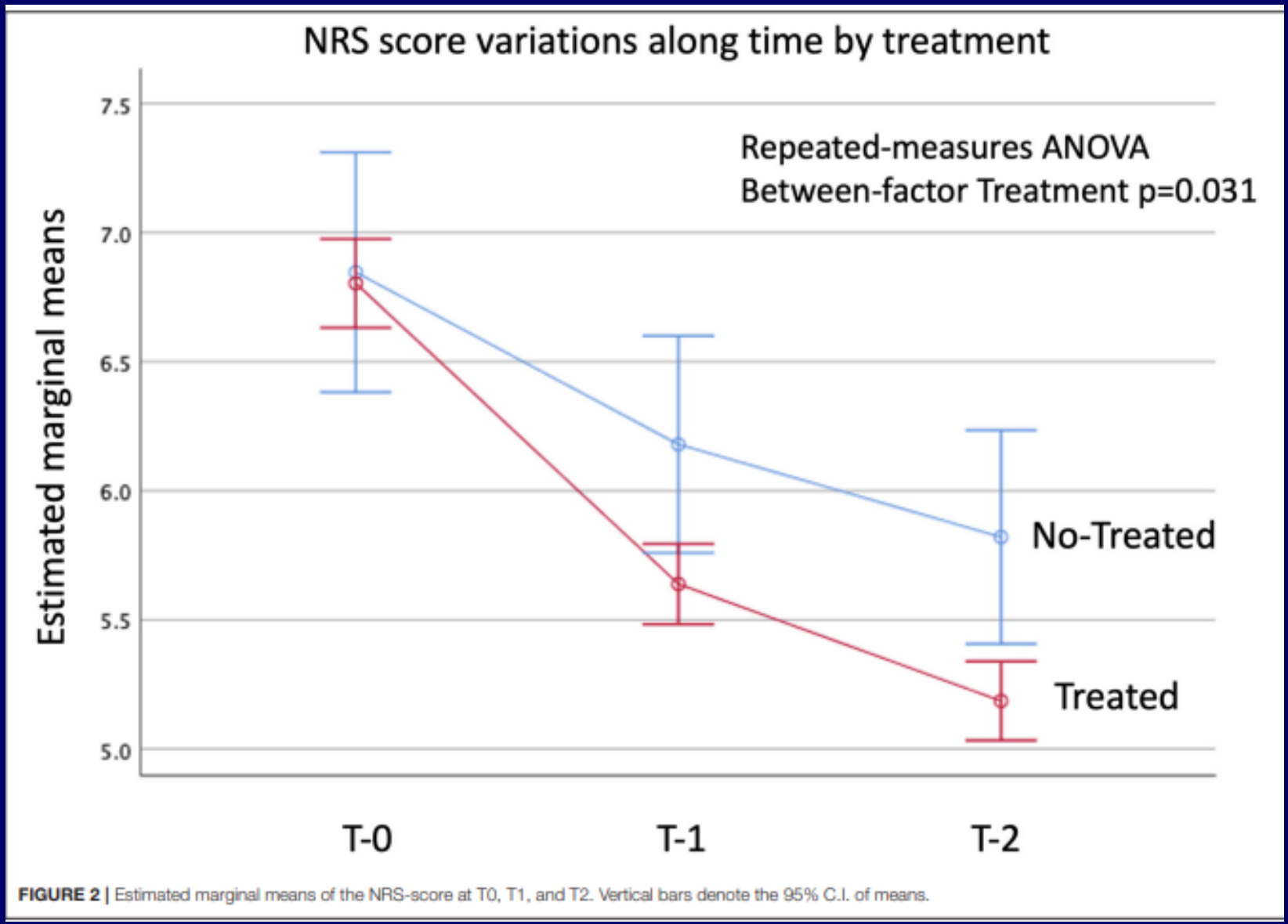

FIGURE 2 | Estimated marginal means of the NRS-score at T0, T1, and T2. Vertical bars denote the 95% C.I. of means.

SUMMARY

- Objective diagnosis of TOS is a continuous challenge due to the wide variety of aspecific symptoms and of differential diagnosis
- Despite several progresses in the diagnostic process in the last 20 y, significant technical issues and controversies still persist
- Clinical suspicion should be confirmed by objective diagnosis and by multisciplinary approach, in order to achieve prompt recognition of the syndrome and a swift start of the treatment for rapid and successful results
- A structured and standardized rehabilitative process should represent the initial treatment for TOS leading to a significant 60-90% symptoms improvement, leaving surgery only for patients who failed to improve after a conservative management program or with refractory or recurrent symptoms
- Albeit encouraging and in line with the literature, our results require confirmation coming from properly designed studies on a larger patient cohort