Scientific Research Publishing

World Journal of Cardiovascular Surgery, 2022, 12, 235-244
https://www.scirp.org/journal/wjcs
ISSN Online: 2164-3210
ISSN Print: 2164-3202

Algorithm for Surgical Decision Making in Patients with Thoracic Outlet Syndrome

Farid Gharagozloo[1,2,3]

[1]Institute for Advanced Thoracic Surgery, Orlando, USA
[2]University of Central Florida College of Medicine, Orlando, USA
[3]University of Central Florida Lake Nona Medical Center, Orlando, USA
Email: Gharagozloo@ThoracicSurgeryInstitute.org

How to cite this paper: Gharagozloo, F. (2022) Algorithm for Surgical Decision Making in Patients with Thoracic Outlet Syndrome. *World Journal of Cardiovascular Surgery*, **12**, 235-244.
https://doi.org/10.4236/wjcs.2022.1210021

Received: September 4, 2022
Accepted: October 8, 2022
Published: October 11, 2022

Abstract

Purpose: Historically the classification of Thoracic Outlet Syndrome (TOS) has been based on symptoms rather than the underlying pathology. Therefore, TOS has been classified into Neurogenic (NTOS), Venous (VTOS or Paget Schroetter Syndrome) and Arterial (ATOS) subgroups. This classification has resulted in confusion among medical practitioners, difficulty in making the diagnosis, and the poor results with surgical intervention. **Methods:** The published papers from PubMed on the newer understanding of the pathogenesis and the surgical treatment of TOS were reviewed. **Results:** More recently TOS has been classified based on the underlying pathologic entity. Based on this classification, patients who are suspected of having TOS should be classified as having 1. Cervical Rib Disease (CRD), or 2. TOS as the result of "Subclavian Vein Compression Syndrome". This classification has resulted in more accurate diagnosis, better patient selection for surgery, and excellent surgical results. This paper outlines the algorithm for making the appropriate diagnosis in patients who present with neurovascular symptoms of the upper extremity and the selection of the appropriate patients for surgery. **Conclusion:** Based on the algorithm for surgical decision making, patients with Cervical Rib Disease should undergo cervical exploration and resection of the pathologic entity which results in compression of the brachial plexus or the subclavian artery in the neck. Patients with Thoracic outlet Syndrome who are found to have extrinsic compression of the subclavian vein by a pathologic tubercle at the sternocostal joint on Multiphasic MRA should undergo robotic first rib resection.

Keywords

Thoracic Outlet Syndrome, Robotic First Rib Resection, Neurogenic TOS, Venous TOS, Arterial TOS, Algorithm

1. Introduction

Since 1956, with the publication of a paper by Peet, a diverse group of patients with symptoms in the shoulder and upper extremity who present with pain, numbness, tingling, and swelling, have been classified under the umbrella of "Thoracic Outlet Syndrome" ("TOS") [1]. Since that report, TOS has been classified based on symptoms, rather than the underlying pathology, with the subgroups of neurogenic (NTOS), Venous (VTOS or Paget Schroetter Syndrome, PSS), and Arterial (ATOS) TOS [2]. Neurogenic TOS accounts for over 95% of the cases, followed by Venous (3% - 5%) and Arterial (1% - 2%) [2]. Less than 1% of patients with the diagnosis of neurogenic TOS are found to have a cervical rib or cervical bands. The majority of patients with symptomatic NTOS have non-specific findings on physical examination, and almost all present with negative nerve conduction velocity (NCV) studies. In the 1980s, Wilbourne asserted that a positive NCV study should be a sine qua non for the diagnosis of neurogenic TOS. Wilbourne further reasoned that patients with an identical clinical picture, but without a positive NCV study, should be considered as having 'Disputed' NTOS ("D"NTOS) [3]. Notwithstanding impressive evidence over the past four decades that NCV demonstrates little sensitivity or specificity in patients with NTOS, some physicians still consider all cases of NTOS as "Disputed". Thus, historically, in patients with "Disputed" NTOS, surgery has been used as a last resort in order to establish the diagnosis. Using surgery as both a diagnostic and therapeutic modality in patients with "D"NTOS has resulted in a high rate of false positive findings, a heterogenous patient population, poor surgical results, and a high rate of complications [4] [5]. Furthermore, this approach has added to the confusion in diagnosing TOS, and surgical decision making.

In the past decade, a number of observations have resulted in reevaluation of the pathogenesis, diagnosis, and treatment of TOS. This communication outlines the results of the newer research which has led to a better understanding of the pathologic cause for TOS, and outlines an algorithm for surgical decision making in these patients.

2. Venous TOS (Paget Schroetter Syndrome PSS)

Examination of the medial aspect of the resected first ribs in patients with PSS has demonstrated the presence of a congenitally malformed bony tubercle which forms a tighter and wider joint at the junction of the first rib and the sternum [6] (**Figure 1**). It has been observed that the wider and less mobile cost-sternal joint "locks" the medial aspect of the first rib into place and results in extrinsic compression by the boney tubercle onto the Subclavian Vein (SV) at its junction with the innominate vein. This abnormal tubercle on the medial aspect of the first rib can be seen on Three-Dimensional (3D) reconstruction of computerized axial tomograms and can be demonstrated on Multiphase Magnetic Resonance Angiography (MRA) with maneuvers as well as dynamic venography (**Figure 2**).

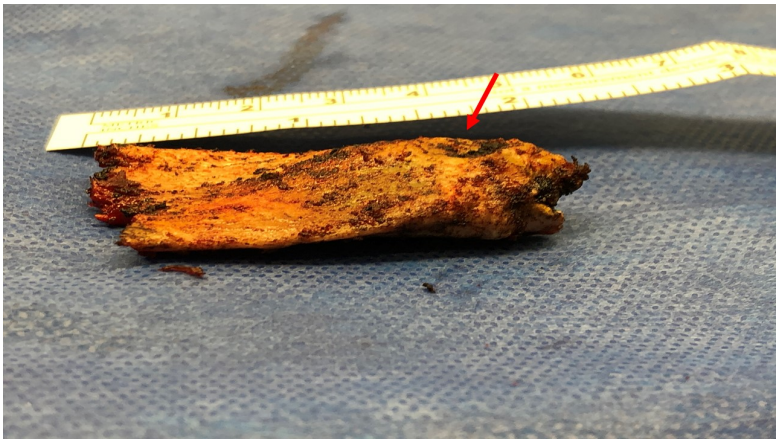

Figure 1. The resected specimen of the "offending" portion of the first rib from a patient with Venous TOS (Paget Schroetter Syndrome) showing the abnormal bony tubercle on the medial aspect of the first rib (arrow) that compresses the subclavian vein at the subclavian-innominate junction.

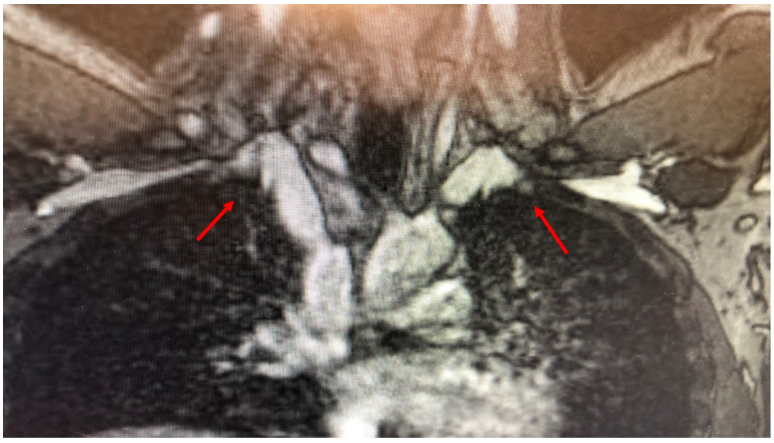

Figure 2. Multiphasic Magnetic resonance angiogram (MRA) with elevation of the left upper extremity shows compression of the subclavian vein by the abnormal bony tubercle in a patient with Paget-Schroetter Syndrome.

Furthermore, these studies have demonstrated that the SV compression increases with elevation of the arm above the shoulder. In patients with PSS, it has been shown that disarticulation of the Costo-Sternal (SC) joint and resection of the "offending portion" of the first rib (portion of the rib medial to the SA) results in decompression of the SV and relief of Paget Schroetter Syndrome with return to full function of the affected upper extremity [7] [8] [9].

3. Neurogenic TOS

Detailed assessment of symptoms in patients who present with VTOS or Paget Schroetter Syndrome (PSS), has revealed a significant history of unrecognized upper extremity neurologic symptoms which predated the clotting of the subclavian vein and onset of Paget Schroetter Syndrome (PSS). The neurologic symptoms were relieved after undergoing resection of the abnormal portion of the

first rib for PSS. Therefore, it has been hypothesized that in a subset of patients, NTOS may be the manifestation of compression of the subclavian vein, and venous congestion resulting in venous ischemia of the upper extremity nerves which lead to diffuse neurologic symptoms unrelated to nerve compression. This hypothesis has been supported by 4 observations: 1. The anatomic features of the first rib in normal individuals was compared to the first ribs resected from patients with NTOS who experienced complete postoperative symptom relief following first rib resection. In patients with NTOS, the first rib exhibited an abnormal bony tubercle immediately lateral to the costo-sternal joint [10] (**Figure 3**). The tubercle was found in place of the subclavian groove which is seen in normal first ribs. In normal individuals, the rib thickness at the subclavian groove was 0.2 cm which represented an indentation of 0.8 cm from the rib surface. It was surmised that at the time of extension of the upper extremity above the shoulder and elevation of the first rib, the presence of this groove prevents compression of the subclavian vein by the rib. On the other hand, in patients with NTOS the thickness of the first rib at the abnormal tubercle was 1.4 cm, which represents a differential elevation of 1.2 cm above the subclavian groove. Given the fact that the subclavian vein has an average diameter of approximately 1 cm in its position over the first rib, it was concluded that the tubercle compressed the subclavian vein at rest, and significantly occluded the vein with extension of the arm above the shoulder. 2. A study of patients with neurogenic TOS who experienced persistent upper extremity pain following first rib resection revealed persistent extrinsic compression of the subclavian innominate junction on dynamic MRA (**Figure 4**). These patients underwent video-assisted exploration of the chest which showed a persistent SC joint despite evidence for prior removal of the first rib. Disarticulation of the cost-sternal joint and removal of the remaining portion of the first rib which bore an abnormal tubercle alleviated the extrinsic compression of the subclavian-innominate vein junction on

Figure 3. The resected specimen of the "offending" portion of the first rib from a patient with "Disputed" Neurogenic TOS showing the abnormal bony tubercle on the medial aspect of the first rib (arrow) that compresses the subclavian vein at the subclavian-innominate junction. This pathologic abnormality is similar to that found in patients with Paget Schroetter Disease.

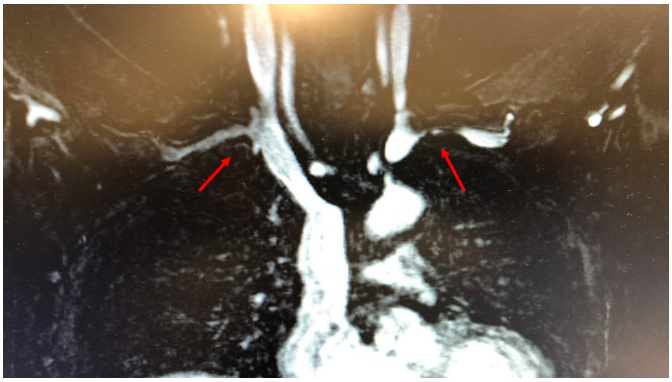

Figure 4. Multiphasic Magnetic resonance angiogram (MRA) in a patient with Disputed Neurogenic TOS. With elevation of the arms, there is bilateral compression (arrows) of the subclavian innominate vein junction.

postoperative dynamic MRA and resulted in relief of Neurogenic symptoms in all patients [11]. 3. In a proof-of-concept study, 97% of patients with NTOS had complete relief of symptoms after robotic resection of the medial aspect of the first rib and disarticulation of the SC joint and relief of the extrinsic boney compression of the SV [12]. 4. Studies have demonstrated that the blood-nerve barrier in the nerve root was more easily broken by venous congestion than by arterial ischemia. Venous congestion may be an essential factor precipitating circulatory disturbance in nerve roots and inducing neurogenic intermittent claudication [13]. Therefore, compression of the SV at its junction with the innominate vein may result in elevation of venous pressure (venous congestion), a decrease in arterial flow, and relative ischemia of the nerves of the upper extremity. Venous congestion of the upper extremity nerves may manifest as pain, tingling, paresthesia and numbness, and varying degrees of neurogenic intermittent claudication depending on the degree and duration of venous compression. Elevation of the extremity above the shoulder may result in greater compression of the SV, further venous congestion, further decrease in arterial flow, greater degree of ischemia of the upper extremity nerves, and exacerbation of symptoms. This phenomenon is demonstrated on dynamic Magnetic Resonance Imaging (MRI) and venography. The pathophysiology of nerve pain in this setting has been likened to symptoms that result from "crossing one leg over the knee". These observations have led to the hypothesis that NTOS may be caused by "Venous Congestion" resulting in secondary neurologic symptoms as opposed to the direct "compressive" of the nerves themselves.

4. Classification of Thoracic Outlet Syndrome

Peet's classification of TOS was based on anatomic rather than symptomatic presentation of the disease. Based on recent studies, in order to decrease confusion and to improve therapeutic results with TOS, it is proposed that the disease should be classified based on the underlying pathologic entity. Acquired and traumatic abnormalities of the clavicle and first rib should be classified separate-

ly. Clearly after the more common and objectively supported diagnoses of conditions that result in neurovascular symptoms of the upper extremity such as cervical spine disease, carpal tunnel disease and nerve entrapment syndromes have been ruled out, there remains a group of patients who are suspected of having TOS. In these patients rather than the historic classification such as arterial, venous, or neurogenic, the more accurate approach from a diagnostic and therapeutic approach may be to classify them as:

4.1. Cervical Rib Disease (CRD)

In these patients an abnormal cervical rib or the associated bands that insert onto the first rib result in compression and displacement of the nerves or vessels in the neck. These patients can present with neurologic or vascular compromise. Patients with CRD can have complications relating to compression of the SA and the brachial plexus secondary to a well-formed cervical rib, or to an incompletely formed first rib, fibrous band associated with a rudimentary cervical rib, or a giant transverse process of C7. Although in the past, these patients have been classified as TOS, separation of these patients into CRD allows for a more precise diagnostic and therapeutic strategy and perhaps more importantly clears the way to a better understanding of diseases that result from anomalies of the first rib.

In patients with neurologic symptoms (previously classified as NTOS) due to variations of Cervical Rib Disease and compression of the brachial plexus, nerve conduction studies and needle electromyography (EMG) may provide useful information [14]. Historically, these cases are classified as true NTOS by most authoritiesbut are quite rare. In these cases, EMG findings that localize to conduction defect with the branches of the brachial plexus are pathognomonic and correlate with the clinical symptoms and the findings on the neurologic exam. The diagnostic algorithm in these patients consists of Sensory and Motor Nerve Conduction studies along with radiographs of either the cervical spine or chest. Some clinicians will perform MRI of the cervical spine to rule out neural foraminal stenosis as a cause of polyradiculopathy.

4.2. Thoracic Outlet Syndrome (TOS)

In these patients an abnormal first rib at the costo-sternal joint results in compression of the SV at the subclavian-innominatejunction [15]. Compression of the vein results in venous hypertension and congestion of the nerves of the upper extremity and resultant neurologic symptoms. With prolonged compression of the subclavian-innominate junction, the vein clots giving rise to PSS. Therefore, patients who have been previously classified as neurogenic and venous TOS represent a variable symptomatic presentation of the same pathologic entity which affects the SV. The term arterial TOS should be abandoned as these patients are better classified under cervical rib disease or under traumatic causes. On the other hand, neurogenic and venous TOS appear to be caused by the same

pathophysiologic process which compresses the SV at its junction with the innominate vein. They represent different clinical manifestations of the same disease and represent the spectrum from pain to venous thrombosis. It is suggested that these conditions may best be classified "subclavian vein compression syndrome".

Historically, no diagnostic test has been well established for the vast majority of patients who present with neurologic symptoms of the upper extremity, without cervical ribs, but who have a normal neurologic exam and nerve conduction studies. Unfortunately, these patients have been classified as having "Disputed" TOS ("D"TOS).

A recent report by the American College of Radiology which addressed guidelines for different radiologic modalities for the diagnosis of TOS concluded that MRA of the chest is one of the most appropriate diagnostic tests for NTOS. In a study that spanned a 10-year period, 157 patients with "D"NTOS underwent robotic resection of the medial aspect of the first rib with disarticulation of the costo-sternal joint. Preoperative Multiphasic MRA showed extrinsic compression of the subclavian-innominate junction at rest and increase of the compression with elevation of the arm in all patients. Intraoperatively the subclavian-innominate junction was decompressed with the disarticulation of the costo-sternal joint (Figure 5). Postoperative Multiphasic MRA showed relief of the extrinsic compression of the subclavian-innominate junction at rest and with arm elevation. Following the procedure, all patients experienced relief of symptoms. The Predictive Value (PV) of preoperative MMRA with maneuvers in determining the success of surgery was 97.5%.

4.3. Algorithm for Surgical Decision Making:

Figure 6 depicts the algorithm for diagnostic strategy and surgical decision making in patients who are suspected as having TOS. Clearly after the more

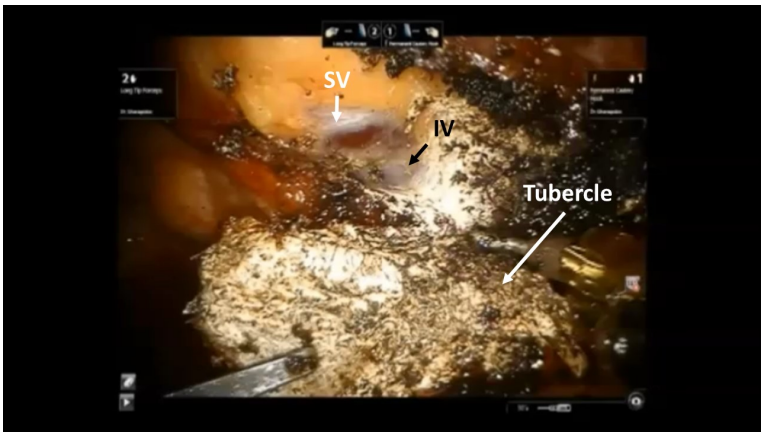

Figure 5. Intraoperative photograph during the robotic resection of the medial right first rib in a patient with Disputed Neurogenic TOS. The abnormal boney tubercle at the costo-steral joint results in compression of the subclavian vein (SV) at its junction with the innominate vein (IV).

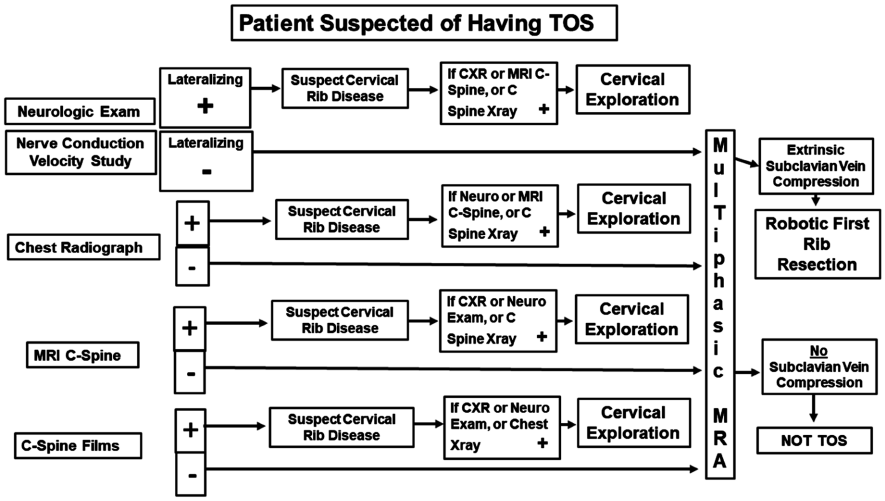

Figure 6. Algorithm for the workup and surgical decision making in patients who are suspected of having Thoracic Outlet Syndrome.

common and objectively supported diagnoses of conditions that result in neurovascular symptoms of the upper extremity such as cervical spine disease, carpal tunnel disease and nerve entrapment syndromes, have been ruled out, there remains a group of patients who are suspected of having TOS. When a patient is suspected of having TOS, they need to undergo:

1) Complete History and Physical examination;
2) Neurologic Examination;
3) Cervical Spine Films;
4) Chest Radiograph;
5) Nerve Conduction Velocity Study (NCV);
6) MRI of the Cervical Spine;
7) Multiphasic MRA of the Thoracic Outlet.

If there are localizing signs for nerve compression or compression of the subclavian artery in the neck, patients should be classified as having Cervical Rib Disease (CRD). Examples of CRD include anomalies of the cervical vertebral bodies which would include an elongated transverse process of a cervical vertebral body, a true cervical rib, or bands extending from the anomalous cervical vertebral body to the first rib. In all these conditions, neurologic exam, NCV, and imagining studies would show the specific entity that can be addressed through a cervical surgical approach.

Patients with Paget Schroetter Disease (VTOS) should undergo lysis of the clot in the subclavian vein (SV), anticoagulation and robotic first rib resection with the purpose of relieving extrinsic compression on the SV.

Patients who do not have objective findings on Neurologic Examination, Cervical Spine Films, Chest Radiograph, Nerve Conduction Velocity Study (NCV), MRI of The Cervical Spine would be classified as "Disputed" NTOS. These patients should undergo Multiphasic MRA with maneuvers. If extrinsic compression of the SV by the abnormally developed medial aspect of the first rib at the

costo-sternal joint is identified, these patients should undergo robotic first rib resection.

5. Conclusion

This paper presents an algorithm for differentiating patients with Cervical Rib Disease from patients with Thoracic Outlet Syndrome based on objective testing in patients who present with neurovascular symptoms of the upper extremity. Patients with Cervical Rib Disease should undergo cervical exploration and resection of the pathologic entity which results in compression of the brachial plexus or the subclavian artery in the neck. Patients with Thoracic outlet Syndrome who are found to have extrinsic compression of the subclavian vein by a pathologic tubercle at the sternocostal joint on Multiphasic MRA should undergo robotic first rib resection.

Conflicts of Interest

The author declares no conflicts of interest regarding the publication of this paper.

References

[1] Peet, R.M. (1956) Thoracic-Outlet Syndrome: Evaluation of a Therapeutic Exercise Program. *Proceedings of the Staff Meetings of the Mayo Clinic*, **31**, 281-287. https://doi.org/10.1155/2014/248163

[2] Freischlag, J. and Orion, K. (2014) Understanding Thoracic Outlet Syndrome. *Scientifca* (*Cairo*), **2014**, 248163.

[3] Wilbourn, A.J. (1999) Thoracic Outlet Syndrome is Overdiagnosed. *Muscle Nerve*, **22**, 130-136. https://doi.org/10.1002/(SICI)1097-4598(199901)22:1<130::AID-MUS22>3.0.CO;2-S

[4] Peek, J., Vos, C., Ünlü, Ç., *et al.* (2016) Outcome of Surgical Treatment for Thoracic Outlet Syndrome: Systematic Review and Meta-Analysis. *Annals of Vascular Surgery*, **40**, 303-326. https://doi.org/10.1016/j.avsg.2016.07.065

[5] Falconer, M.A. and Li, F.W. (1962) Resection of the First Rib in Costoclavicular Compression of the Brachial Plexus. *Lancet*, **1**, 59-63. https://doi.org/10.1016/S0140-6736(62)91716-6

[6] Gharagozloo, F., Meyer, M., Tempesta, B., *et al.* (2012) Proposed Pathogenesis of Paget-Schroetter Disease: Impingement of the Subclavian Vein by a Congenitally Malformed Bony Tubercle on the First Rib. *Journal of Clinical Pathology*, **65**, 262-266. https://doi.org/10.1136/jclinpath-2011-200479

[7] Gkikas, A., Lampridis, S., Patrini, D., *et al.* (2022) Thoracic Outlet Syndrome:Single Center Experience on Robotic Assisted First Rib Resection and Literature Review. *Frontiers in Surgery*, **9**, Article ID: 848972. https://doi.org/10.3389/fsurg.2022.848972

[8] Gharagozloo, F., Meyer, M., *et al.* (2012) Robotic Enbloc First-Rib Resection for Paget-Schroetter Disease, a Form of Thoracic Outlet Syndrome: Tech-nique and Initial Results. *Innovations*, **1**, 39-44. https://doi.org/10.1097/imi.0b013e3182542ab3

[9] Gharagozloo, F., Meyer, M., Tempesta, B., *et al.* (2019) Robotic Transthoracic First Rib Resection for Paget-Schroetter Syndrome. *European Journal of Cardio-Thoracic*

Surgery, **55**, 434-439. https://doi.org/10.1093/ejcts/ezy275

[10] Gharagozloo, F. and Meyer, M. (2022) Proposed Pathogenesis for a Subgroup of Patients with Neurogenic Thoracic Outlet Syndrome. *Journal of Clinical Surgery and Research*, **3**, No. 4. https://doi.org/10.31579/2768-2757/045

[11] Gharagozloo, F., Werden, S., Meyer, M., *et al.* (2022) Historic Basis for the New Developments in the Diagnosis and Treatment of Thoracic Outlet Syndrome (TOS). *Clinical Surgery Journal*, **1**, 1-16.

[12] Gharagozloo, F. and Meyer, M. (2022) Robotic Transthoracic First Rib Resection for Neurogenic Thoracic Outlet Syndrome. *World Journal of Cardiovascular Surgery*, **12**, 1-11. https://doi.org/10.4236/wjcs.2022.121001

[13] Kobayashi, S., Takeno, K., Miyazaki, T., *et al.* (2008) Effects of Arterial Ischemia and Venous Congestion on the Lumbar Nerve Root in Dogs. *Journal of Orthopaedic Research*, **11**, 1533-1540. https://doi.org/10.1002/jor.20696

[14] Ferrante, M.A. and Wilbourn, A.J. (1995) The Utility of Various Sensory Nerve Conduction Responses in Assessing Brachial Plexopathies. *Muscle & Nerve*, **18**, 879-889. https://doi.org/10.1002/mus.880180813

[15] Gharagozloo, F. (2022) Reevaluating the Pathogenesis and Classification of Thoracic Outlet Syndrome. Academia Letters, Article 5408. https://doi.org/10.20935/AL5408