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Is there something new in the diagnosis and treatment of TOS and Paget-Schroetter syndrome?

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Abstract

Thoracic outlet syndrome is a syndrome of pathologic neurologic and vascular symptoms involving the upper limb. Its most serious complication, Paget-Schroetter syndrome, is a major clinical problem, posing a significant diagnostic and therapeutic challenge to specialists not only in angiology and vascular surgery but also in neurology and physiotherapy. There is a need for in-depth examination and differentiation of patients with suspected compression syndrome. We present a review paper covering the diagnosis and treatment of thoracic outlet syndrome.

Key words: Paget-Schroetter syndrome, thoracic outlet syndrome, venous thrombosis, compression syndrome

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Introduction

Thoracic outlet syndrome (TOS) is a syndrome of pathologic neurologic and vascular symptoms involving the upper limb [1–3]. The most common cause is compression of the brachial plexus region and the upper limb's vascular bundle in this area, including the subclavian and axillary arteries and the subclavian vein [1, 4, 5]. Compression and damage most often occur at the neurovascular bundle between the cervical spine and the upper limb's axillary region [5, 6]. The causes of compression syndrome include anatomical conditions (congenital anomalies and past trauma), as well as postural abnormalities in the muscular structures and ligamentous apparatus of the upper thoracic orifice [5–8]. The literature emphasizes among the congenital anomalies causing compression of neurovascular structures such as defects as an additional cervical rib, residual first rib, first bicuspid rib, fibromuscular pathologies in the triangle of inclined muscles, displacement of inclined muscle attachments or their adhesions [4, 5, 9, 10]. Authors of publications covering this issue emphasize that, to a large extent, upper thoracic compression syndrome may develop in patients with abnormal posture during work, with the so-called physiological dropping of the shoulder girdle [7, 8]. However, it is also a phenomenon affecting athletes, especially those practicing sports requiring repeated use of specific movements with excessive upper limb strength (e.g. during gym exercises) [11]. There is also a large group of patients with

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excessive hypertrophy of the shoulder girdle muscles, such as bodybuilders, construction workers, or miners [11]. Trauma, especially to the clavicle and first rib, and less commonly, dislocations of the humeral head, also play a role in developing compression symptoms in the upper thoracic orifice [2, 4]. The importance of stress as a predisposing factor contributing factor in the weakening and decreased capacity of structures within the upper thoracic orifice has received increasing attention in the literature [4]. TOS syndrome is estimated to occur in approximately 0.3–8.0% of the population [10]. It is more repeatedly seen in women, and the average age of patients developing symptoms of the syndrome is between 30–40 years [1, 4]. Authors of publications related to this issue emphasize the variability of the symptoms of compression syndrome. It is emphasized that the manifestations of the upper thoracic compression syndrome depend primarily on the degree of compression applied to the brachial plexus and the choroid bundle [6]. They are usually classified in the literature into three primary groups of symptoms. In the first group, symptoms of the so-called venous syndrome are noted, which affect 3–5% of patients and result from compression exerted on the subclavian or axillary vein between the clavicle and the first rib [2, 5, 12]. This most severe complication of the venous component of compression has been described as the so-called Paget-Schroetter syndrome, resulting in thrombosis of the upper limb veins at the compression site, which can lead to life-threatening thromboembolic complications, including pulmonary embolism and even death [2, 3]. Paget-Schroetter syndrome usually affects young people and usually results from prolonged physical exertion involving the upper limbs. Among the predominant symptoms are dull pain and a feeling of heaviness of the limb, especially aggravated in the supine position due to prolonged fixed position of the upper limb, mainly during sleep [2, 3]. In the clinical presentation, purplish-red discoloration and increasing swelling of the limb are observed over time [1–3]. During the disease development, significantly dilated venous vessels of the upper extremity, chest wall, and neck are observed [3]. In the case of the so-called arterial syndrome, which affects 1–2% of patients with TOS, an acute or chronic disease course is noted [2]. The arterial syndrome symptoms often result from local irritation of the arterial wall against the base of the first rib, which leads to arterial fibrosis and sometimes to the development of an aneurysm [13–15]. In the acute state of the arterial syndrome, symptoms of acute limb ischemia are usually observed by thrombotic and embolic mechanisms within the artery and microemboli in the small vessels of distal parts of the upper limbs [16]. In the chronic phase, a cold sensation of the hand skin is observed, often Raynaud's sign and symptoms analogous to those observed in chronic ischemia, such as intermittent claudication, increasing trophic changes in fingers, and with increasing ischemia, even subungual ulceration and necrosis [2, 3]. The most numerous group of symptoms resulting from upper thoracic compression syndrome are those of neurogenic syndrome, which are reported in approximately 95% of patients with TOS and result from compression or injury to the brachial plexus [6]. As noted in the literature, patients with symptoms of neurogenic syndrome typically report complaints of cervical spine or shoulder pain, upper extremity paresthesias, numbness, and tingling in the entire arm or forearm and hand [1, 6]. Over time, motor and muscle strength disorders in the forearm and hand may also occur. In the case of compression syndrome of the upper thoracic aperture and its most serious complication, Paget-Schroetter syndrome, the diagnosis of the disease is based on the clinical history, the so-called provocative tests, and extensive imaging diagnostics including both non-invasive and invasive diagnostic procedures used in vascular surgery and angiology [2, 3, 17].

Diagnosis of TOS and Paget-Schroetter syndrome

Diagnosis of TOS and its complication Paget-Schroetter syndrome is based on medical history, provocative tests, diagnostic ultrasound, noninvasive and invasive radiological procedures [12, 18]. The literature emphasizes the significance of provocative tests such as Adson's test, Falconer and Weddel's test, Wright's hyperabduction test, AER (abduction-external-rotation) test, based mainly on the intensification of existing compression of structures of the upper limb neurovascular bundle [12, 18, 19]. As noted in the literature, provocative tests have limited specificity for these syndromes and are characterized by a high rate of false positives. One of the most frequently performed provocative tests is the Adson's test, which evaluates a segment of the subclavian artery in the triangle of the oblique muscle [12]. This test, described by American neurosurgeon Alfred Washington Adson, relies on the disappearance of the radial artery pulse when the subject turns his or her head toward the diseased side after taking a deep breath. One of the widely used clinical provocative tests is the hyperabduction test, which allows assessment of the subclavian artery at the pectoralis minor muscle attachment [4, 18]. This test is often used in conjunction with flow assessment of the subclavian vein and artery by ultrasound and arteriography. As emphasized in the literature and clinical studies, this test is currently not recommended due to a high rate of false-positive

results (about 50%) [19]. Clinical studies emphasize the significance of imaging in the diagnosis of TOS and its complications [19]. Special attention is given to the use of radiological examinations, including traditional X-rays of the chest and cervical spine and shoulder joints, which allow the diagnosis of possible anatomical and functional changes of the skeleton based on which the compression syndrome develops. Of particular importance in patients with TOS is ultrasound diagnostics, especially the dual ultrasound imaging method [2, 18, 19]. The literature emphasizes the high sensitivity and specificity of ultrasonography in diagnosing flow abnormalities in the upper extremities' venous and arterial vessels. In addition to anatomical changes and arterial complications, it allows functional assessment of the vascular system [18]. Nevertheless, the anatomy of the upper thoracic aperture region may pose some limitations to ultrasound evaluation, especially in the venous system, especially when thrombotic complications of upper thoracic aperture syndrome are suspected. It is recommended to extend the diagnosis with radiological studies using phlebography, arteriography, or phlebography and computed tomography arteriography in case of diagnostic doubt [1–3, 18]. Since venous thrombosis of the upper limb is the most severe complication of TOS, there are critical clinical reasons in the literature for using phlebography as the primary method of diagnosis [16, 20]. Phlebography is recommended in all cases of acute thrombosis of the subclavian vein, with attention paid to the fact that the examination should be performed using the Seldinger method [20]. It is crucial that phlebography, apart from its diagnostic value in diagnosing thrombotic complications of TOS, allows local thrombolysis and possible endovascular plastic surgery [21]. The authors of the studies are unanimous in stating that phlebography is one of the methods to determine the cause of the compression syndrome [2]. In the literature, the diagnosis of TOS is considered an indication for surgical treatment, which consists of resection of the first rib and scalenectomy [18]. Nevertheless, it is strongly stated that of great value in treating patients with this condition is the implementation as soon as possible of physical therapy treatment aimed at relieving the vascular and nerve bundle of the upper thoracic aperture region. [17, 21–23].

The treatment of TOS and Paget-Schroetter syndrome

Authors of clinical studies discussing the treatment of patients with TOS syndrome emphasize the importance of physical therapy, including rehabilitation, consisting of physical therapy and kinesitherapy [19, 24, 25]. The main goal of physical therapy in treating this condition is to reduce pain and increase muscle tone in the shoulder girdle and cervical spine. Physical therapy usually depends on the type of compression syndrome, the type of pain, and the patient's tolerance to the treatments. As emphasized in the literature, physical therapy is used to prepare the patient for kinesitherapy treatment, which may be successfully applied simultaneously [7, 24, 25]. Kinesitherapy in patients with TOS syndrome should be strictly personalized, subject to appropriate individual selection. Before initiating kinesitherapy treatment, the patient's capacity should be considered; after the physiotherapeutic examination, the type of therapeutic techniques and exercises, their starting position, and degree of intensity are selected [24]. The authors emphasize that kinesitherapy is one of the essential elements of the combined treatment of patients with compression syndrome [21]. Attention is drawn to the significant acceleration of repair processes, protects against the development of abnormal compensatory schemes, prevents secondary changes in the musculoskeletal system [25]. Particularly noteworthy is that in the applied assessment of the mobility of the joints of the upper thoracic orifice, segments are differentiated into hypomobile and hypermobile ones. Segments characterized by low mobility (hypomobile) should be mobilized in order to maintain maximum motor harmony. In segments with excessive mobility (hypermobility), treatments are aimed at stabilizing them. The stability of segments of the upper thoracic aperture, especially the cervical spine and shoulder girdle, depends on the functional and stable position of the scapula. The imbalance of muscle tone in the scapular region (weakening of the inferior part of the serrates anterior muscle and inferior part of the trapezius muscle, and excessive tension of the pectoralis minor and rhomboid muscles) leads to protraction of the scapula and distension of its lower angle from the thorax. A common postural problem in TOS is the presence of excessive forward head extension leading to overuse of the sternocleidomastoid muscles [7, 25]. When the superficial Sternocleidomastoid muscles are overactive, there may be a tendency to underactive the deep cervical flexors, leading to cervical dysfunction. Various soft tissue therapy techniques are used to reduce excessive muscle tension. However, what is crucial for physiotherapeutic management is the patient's education on ergonomics, especially during heavy work duties. The literature emphasizes that in patients with neurological symptoms in the compression syndrome, especially despite the intensification of conservative management, or especially in patients with vascular complications with particular emphasis on potential thromboembolic complications, surgical intervention should be considered [6, 18]. For potential arterial

complications, a vascular reconstruction procedure is usually performed [8]. As emphasized in the literature, occlusion of the arterial lumen due to compression by the cervical rib is usually eliminated by removing the first rib [10]. Surgical eligibility for surgical treatment in upper thoracic orifice compression syndrome is usually based on a broad diagnostic spectrum including clinical trials, noninvasive studies such as chest radiographs, cervical spine radiographs, computed tomography, and magnetic resonance imaging, especially with contrast and evaluation of the results of invasive imaging such as phlebography and arteriography [21]. As underlined extensively in the literature, the surgical treatment of choice mainly involves resection of the first rib, often combined with surgery to remove bony or fibromuscular pathologies [5, 10, 12, 19]. Surgery is usually assumed to be necessary when vascular complications, both venous and arterial, are confirmed on imaging examinations [5, 18]. In the case of the venous complication of TOS, such as Paget-Schroetter syndrome, it is believed that the effects of surgical treatment of venous compression allow stenting to be omitted, and decompression of the subclavian vein prevents recurrent occlusion or stenosis of the vessel in most cases [26, 27]. As pointed out in the literature, the preferred and appropriate method of revascularization in the subclavian vein is the use of thrombolytic therapy, noting that thrombectomy is rarely performed, potentially resulting in recurrent thromboembolism [21, 27].

Summary

Thoracic outlet syndrome, including its most serious complication, Paget-Schroetter syndrome, is a major clinical problem, posing a significant diagnostic and therapeutic challenge to specialists not only in angiology and vascular surgery but also in neurology and physiotherapy. It is particularly noted that patients complaining of TOS symptoms and diagnosed complications are mainly young, physically, and professionally active people. Thus, the symptoms of compression syndrome may cause a significant reduction in the quality of life and lead to life-threatening complications in extreme cases. As emphasized in the literature, there is a need for in-depth examination and differentiation of patients with suspected compression syndrome. The multitude of currently existing diagnostic methods greatly contributes to the improvement of the diagnosis and the incorporation of appropriate therapeutic management. The variety of symptoms, their varying nature and severity accompanying thoracic outlet syndrome, and potentially life-threatening complications such as Paget-Schroetter syndrome, among others, usually require a multidisciplinary evaluation of patients, their potential risk factors, and their possible response to the therapeutic management used.

Conflict of interest

None.

References:

1. Adamski S. Zespoły naczyniowo-nerwowe. In: Rykowski H. ed. Choroby naczyń. PZWL, Warszawa 1990: 442–454.
2. Sieroń A, Cierpka L, Rybak Z, Stanek A. Podręcznik angiologii. Alfa-medica Press 2009.
3. Craeger MA, Dzau V, Loscalzo J. Choroby naczyń. Elsevier 2006: Elsevier.
4. Mackinnon SE, Novak CB. Thoracic outlet syndrome. Curr Probl Surg. 2002; 39(11): 1070–1145, doi: 10.1067/msg.2002.127926, indexed in Pubmed: 12407366.
5. Mackinnon SE, Novak CB. Thoracic outlet syndrome. Curr Probl Surg. 2002; 39(11): 1070–1145, doi: 10.1067/msg.2002.127926, indexed in Pubmed: 12407366.
6. Davidovic L, Kostic D, Jakovljevic N, et al. Vascular thoracic outlet syndrome. World J Surg. 2003; 27(5): 545–550, doi: 10.1007/s00268-003-6808-z.
7. Novak C. Thoracic outlet syndrome. Clinics in Plastic Surgery. 2003; 30(2): 175–188, doi: 10.1016/s0094-1298(02)00095-0.
8. Schneider D, Curry T, Eichler C, et al. Percutaneous Mechanical Thrombectomy for the Management of Venous Thoracic Outlet Syndrome. Journal of Endovascular Therapy. 2003; 10(2): 336–340, doi: 10.1583/1545-1550(2003)010<0336:pmtftm>2.0.co;2.
9. Edwards DP, Mulkern E, Raja AN, et al. Trans-axillary first rib excision for thoracic outlet syndrome. J R Coll Surg Edinb. 1999; 44(6): 362–365, indexed in Pubmed: 10612957.
10. Hempel GK, Shutze WP, Anderson JF, et al. 770 consecutive supraclavicular first rib resections for thoracic outlet syndrome. Ann Vasc Surg. 1996; 10(5): 456–463, doi: 10.1007/BF02000592, indexed in Pubmed: 8905065.
11. Koffler KM, Kelly JD. Neurovascular trauma in athletes. Orthop Clin North Am. 2002; 33(3): 523–34, vi, doi: 10.1016/s0030-5898(01)00004-9, indexed in Pubmed: 12483948.
12. Pupka A, Szyber P. Zespół uciskowy górnego otworu klatki piersiowej — diagnostyka i leczenie. e-Anatomy. 2021; 8: 16–21.
13. Davidović L, Marković D, Pejkić S, et al. Subclavian Artery Aneurysms. Asian Journal of Surgery. 2003; 26(1): 7–11, doi: 10.1016/s1015-9584(09)60206-2.
14. Kostewicz W. Tętniaki tętnicy podobojczykowej. Medycyna 2000. 1992; 3: 10–11.
15. Szydłowski Z., Szyber P., Pupka A. i wsp. Tętniaki tętnicy podobojczykowej w przebiegu zespołu uciskowego górnego otworu klatki piersiowej. Pamiętnik XXIV Zjazdu Sekcji Chir. Kl. Piersiowej, Serca i Naczyń TchP. Poznań 1992: 311.
16. Pupka A., Szydłowski Z., Janczak D. i wsp. Zakrzepica żyły podobojczykowej i pachowej w przebiegu zespołu uciskowego górnego otworu klatki piersiowej. Flebolimfologia. Wydanie specjalne. II Kongres Pol. Tow. Flebol. Wrocław 1997: 16–18.
17. Angle N, Gelabert HA, Farooq MM, et al. Safety and efficacy of early surgical decompression of the thoracic outlet for Pag-

et-Schroetter syndrome. Ann Vasc Surg. 2001; 15(1): 37–42, doi: 10.1007/s100160010017, indexed in Pubmed: 11221942.
18. Dunant JH. Diagnosis of thoracic outlet syndrome and indications for surgery. Vasa. 1987; 16(4): 345–348, indexed in Pubmed: 3424952.
19. Pupka A. Ocena wyników badań diagnostycznych i leczenia operacyjnego w zespole uciskowym górnego otworu klatki piersiowej. Acta Angiologica. 1995; 1: 51–59.
20. Feugier P, Aleksic I, Salari R, et al. Long-term results of venous revascularization for Paget-Schroetter syndrome in athletes. Ann Vasc Surg. 2001; 15(2): 212–218, doi: 10.1007/s100160010043, indexed in Pubmed: 11265086.
21. Schmacht DC, Back MR, Novotney ML, et al. Primary axillary-subclavian venous thrombosis: is aggressive surgical intervention justified? Vasc Surg. 2001; 35(5): 353–359, doi: 10.1177/153857440103500505, indexed in Pubmed: 11565039.
22. Lokanathan R, Salvian AJ, Chen JC, et al. Outcome after thrombolysis and selective thoracic outlet decompression for primary axillary vein thrombosis. J Vasc Surg. 2001; 33(4): 783–788, doi: 10.1067/mva.2001.112708, indexed in Pubmed: 11296332.
23. Novak CB, Collins ED, Mackinnon SE. Outcome following conservative management of thoracic outlet syndrome. J Hand Surg Am. 1995; 20(4): 542–548, doi: 10.1016/S0363-5023(05)80264-3, indexed in Pubmed: 7594276.
24. Aligne C, Barral X. Rehabilitation of patients with thoracic outlet syndrome. Ann Vasc Surg. 1992; 6(4): 381–389, doi: 10.1007/BF02008798, indexed in Pubmed: 1390029.
25. Novak CB. Conservative management of thoracic outlet syndrome. Chest Surg Clin N Am. 1999; 9: 747–760.
26. Reineberg PB, Chang B, Darling III RC. Long-term results in patients treated with thrombolysis, thoracic inlet decompres- sion, and subclavian vein stenting for Paget-Schroetter syndrome. J Vasc Surg . 2001; 33: 100–105, doi: 10.1201/9780429434020-47.
27. Urschel H, Patel A. Paget-Schroetter syndrome therapy: failure of intravenous stents. The Annals of Thoracic Surgery. 2003; 75(6): 1693–1696, doi: 10.1016/s0003-4975(03)00116-4.