

# Bilateral thoracic outlet syndrome: An uncommon presentation of a rare condition in children

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## Abstract

We report an adolescent girl who had left-sided neurogenic thoracic outlet syndrome (TOS) due to impingement of the scalenus anterior muscle with bilateral changes on nerve conduction studies and responded well to surgical decompression. A 13-year-old Caucasian girl presented with intermittent pain, swelling, erythema, tingling and numbness of the palmar aspect of her left hand. Nerve conduction studies revealed bilateral ulnar sensory and motor conduction abnormalities, suggesting early compressive neuropathy in the asymptomatic arm as well. She underwent surgical exploration when it was noted that the scalenus anterior itself was impinging on the brachial plexus. She had a good clinical response to scalenectomy. The diagnosis of neurogenic TOS remains difficult as no single test has been accepted as a gold standard. But, once diagnosed using clinical symptoms, nerve conduction studies, electromyography and radiological investigations, it is a treatable condition with good prognosis.

## Key Words

Disease management, scalenus anticus syndrome, thoracic outlet syndrome

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## Introduction

Thoracic outlet syndrome (TOS) was first described by Rob and Standevan as a group of entities related to compression from the anterior scalene muscle, cervical ribs and other compressive structures.<sup>[1]</sup> The incidence of TOS has been reported to be approximately 0.3–2% of the general population between the ages of 25 and 40 years,<sup>[2]</sup> and is much rarer in the younger population. The symptoms are varied and nonspecific; therefore, the condition can be easily missed. Unless there is evidence of significant motor deficit, subclavian artery compression or debilitating sensory symptoms, the treatment should be kept conservative for as long as possible by a physical therapy program. In refractory cases, the patients will be helped by decompressive operation of the thoracic outlet.

We report an adolescent girl who suffered from a left-sided neurogenic TOS due to impingement of the scalenus anterior muscle and responded well to surgical decompression.

Interestingly, she also had abnormal nerve conduction changes in the right arm suggesting early compressive neuropathy in the asymptomatic arm. This is the first case of subclinical bilateral thoracic outlet syndrome due to scalenus anterior muscle impingement reported in children.

## Case Report

A 13-year-old Caucasian girl, who was otherwise healthy, presented with intermittent swelling, erythema, tingling and numbness of the palmar aspect of her left hand. The nature of the pain was described as dull and shooting. These symptoms lasted for 7–10 days, even though regular analgesics were used.

On examination, the patient was found to be in excellent physical condition. Left hand pulses were easily felt. She had a slight functional limitation of her left hand grip due to pain and stiffness. The left hand was slightly cold to touch compared with the right hand. On examination, the patient was found to be in excellent physical condition. A positive Tinel test and elevated arm test was noted in the clinically affected arm. There was no asymmetry of clavicles noted. There was no muscle atrophy identified.

In view of these symptoms, neurovascular TOS was considered. X-ray of her cervical spine revealed an anomalous first rib that was fused to the second rib [Figure 1]. Magnetic resonance imaging of her brain, cervical spine and brachial plexus

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was normal. She also had a nerve conduction study, which interestingly revealed bilateral ulnar sensory and motor conduction abnormalities as well as early bilateral median sensory nerve conduction changes.

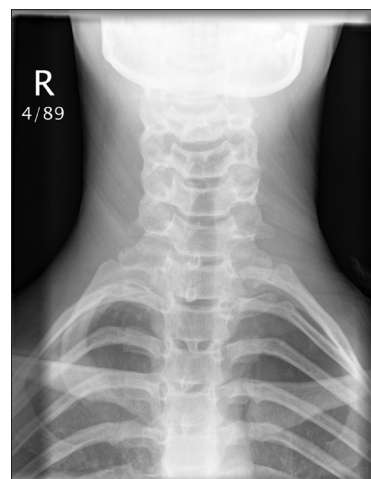
In view of the neuropathic nature of the intermittent pain, she was started on amitriptyline and gabapentin. This had a variable effect on her pain. Following a surgical opinion, a left-sided supraclavicular approach for exploration was undertaken. During the surgery, there was no apparent bony structure compressing the brachial plexus, nor was there any fibrous band. On moving the arm around, it was evident that the brachial plexus was entirely free. The scalenus anterior itself was impinging on the brachial plexus; hence, she underwent left anterior scalenectomy. She showed good response following the surgery. This was evident from the reduction of her symptoms of pain, swelling and tingling. Postoperative sensory and motor nerve conduction studies in the median and ulnar nerves were normal.

## Discussion

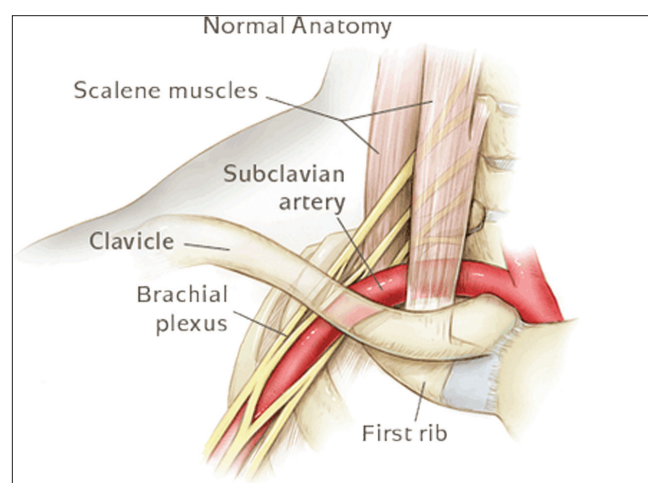
The superior thoracic aperture is referred anatomically as the thoracic inlet and clinically as the thoracic outlet. The clinical entity of TOS is related to this anatomical space. The thoracic inlet is bounded by the first thoracic vertebra posteriorly, the first pair of ribs laterally, the costal cartilage of the first rib and the superior border of the manubrium anteriorly. The inferior thoracic aperture is much larger than the superior thoracic aperture. It is bounded by the edges of the lowest ribs and is closed by the diaphragm that separates the thoracic and abdominal cavities. The thoracic outlet contains three important structures: the brachial plexus, the subclavian artery and the subclavian vein [Figure 2]. As they pass into the upper extremity, these structures run through three important spaces: the interscalene triangle, the costoclavicular space and the subpectoral space. Compression can occur in any of these spaces secondary to trauma or a structural malformation.

TOS is usually categorized in three types: vascular, neurogenic and disputed neurogenic.<sup>[3]</sup> A bony abnormality or soft tissue problem usually plays the etiological role. The bony abnormality could be a cervical rib, abnormal first rib, long transverse process of 7<sup>th</sup> cervical vertebrae or fractures of clavicle. The soft tissue pathologies implicated with TOS are either abnormal fibrous bands and ligaments or congenital/acquired alterations of scalenus anterior muscle. Rapid growth in adolescence could also precipitate the symptomatology as the anatomy of the thoracic apertures is constantly changing.<sup>[4]</sup> Symptoms similar to neurogenic TOS could be seen in Parsonage Turner syndrome, in which inflammation of the brachial plexus leads to shoulder and arm pain followed by weakness and/or numbness.

TOS has been well described in the adult population, but is less frequently reported in those in the first two decades of life. Maru *et al.*<sup>[5]</sup> in their retrospective study identified 12 patients with a mean age of 16.8 years who presented with TOS. Thirty-eight percent of these children presented with neurogenic symptoms, whereas the other 62% presented with either ischemic or venous symptoms. In the adult population, 94–97%



**Figure 1:** X-ray of the cervical spine showing the anomalous 1st left rib fused with the 2nd left rib projected over the clavicle in the image



**Figure 2:** Normal anatomy of the thoracic outlet (adapted from uvahealth.com)

presented with neurogenic symptoms.<sup>[6-8]</sup> Our patient presented with significant neurogenic symptoms and, although the cervical X-ray revealed fusion of the first and second ribs, this was not contributing to the symptoms. On surgical exploration, the scalenus anterior muscle impingement was found to be the contributory mechanism toward the neurogenic symptoms. We could only hypothesize that the nerve conduction study abnormality in the asymptomatic right arm was an early sign of evolving nerve compression. Therefore, the rapid growth in our adolescent patient leading to changing anatomy of the thoracic outlet could be the underlying cause of the bilateral nerve conduction changes.

In the majority of cases, TOS remains a diagnosis of exclusion. The main differential diagnoses include a cervical radiculopathy by soft cervical disc herniation and a distal compression neuropathy. Syringomyelia, Pancoast's tumor and brachial plexitis are rarer pathologies that should be kept in mind.

The diagnosis of neurogenic TOS remains difficult as there

is no single test that has been accepted as a gold standard to establish the diagnosis. In the younger population, this may be even more difficult, as the pediatricians are less likely to consider this disorder as the cause of the child's symptoms. But, once diagnosed using clinical symptoms, nerve conduction studies, electromyography and radiological investigations, it is a treatable condition with good prognosis. Neurogenic TOS should be considered in children even if they present with bilateral symptoms.

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