Thoracic Outlet Syndrome in Children and Young Adults

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Abstract

Objectives: Thoracic outlet syndrome has been well described in the population between 25 and 40 years of age, and is less frequently reported in those in the first two decades of life. The objective of this study was to review results with onset of TOS in the first two decades of life to determine type of presentation and outcomes from surgical intervention.

Methods and materials: Charts of all patients in the first two decades of life, operated on for TOS between 1994 and 2006 were reviewed with follow-up by clinic visit and phone survey to assess the patients' current level of activity and relief from symptoms.

Results: Twelve patients were identified (13 operations), with a mean age of 16.8 years. Acute ischemic symptoms were the initial presentation for 38%, venous TOS in 24%, and neurogenic symptoms in 38%. All patients had symptom relief with surgery with a mean time to resolution of 10.9 weeks. All patients remained symptom free or improved at follow-up.

Conclusions: Vascular TOS is much more common in TOS presenting in the first two decades of life. Surgical intervention for TOS in this population results in long-lasting symptom relief and should be considered for all subtypes of patients.

Introduction

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,1 and is much rarer in the younger population. In the adult experience, 95% of patients present with neurological symptoms and only 5% with vascular symptoms.2–4

A simple Medline search generates greater than 1700 articles on TOS in adults, but only 10 are generated in a search for pediatric TOS. All of these are case reports or descriptions of anatomic abnormalities. There is no good data published to determine the pattern of disease and outcomes from intervention of early-onset TOS. The
objective of our study was to evaluate our experience with early-onset TOS.

**Materials and Methods**

**Data collection**

A retrospective chart review of all patients who presented in a university health system in the first two decades of life and underwent surgery for TOS from 1994 to 2007 was conducted. Data included age of presentation, symptoms, delay to treatment from initial symptoms, tests performed (PPG, US, CT, MRI, EMG), operation, pre- and postoperative activity status, rehabilitation, and complications. IRB approval was obtained for the study prior to contacting patients. Follow-up was obtained from chart and direct patient contact with questionnaire to assess the patient’s current activity status. The survey included questions regarding current activity status, rehabilitation, complications, symptom resolution, pain medication, and overall condition.

**Operative approach**

All patients were evaluated by vascular surgeons both preoperatively and for follow-up. Testing and therapy depended on the presenting symptoms. Those with arterial or venous symptoms underwent angiography or venography when indicated, and were operated on without a lengthy trial of medical management. Arterial TOS (ATOS) was diagnosed if the patient had evidence of embolic disease, arterial aneurysmal formation, or pain with use of the arm associated with positions that occluded flow through the subclavian artery, confirmed with both physical exam and arteriography. Venous TOS (VTOS) was diagnosed if the patient presented with deep vein thrombosis of the axillo-subclavian vein in the area of the thoracic outlet. Neurogenic TOS (NTOS) was diagnosed in those with appropriate history and physical findings on provocative tests, and exclusion of other etiologies for their upper extremity symptoms by cervical spine films, CT, and EMG. Provocative tests included Adson’s, Wrights, Costoclavicular or military position, Roos or EAST test, and hyperabduction of both upper extremities with attention to symptom reproduction, development of pulse changes and bruits. Those with NTOS underwent more extensive evaluation and medical management prior to surgical intervention. However, many of these patients had frequently already undergone extensive testing and attempts at appropriate medical management prior to referral to the vascular surgeon. The standard operative approach utilized was a supraclavicular approach with anterior scalene muscle resection, brachial plexus neurolysis for neurogenic TOS, and segmental first rib resection. In addition, three cervical ribs were resected and a subclavian aneurysm resection was performed. Additional compressive muscle slips or fibrous bands were resected when present. All of the patients operated on for venous TOS initially underwent successful thrombolytic therapy prior to elective surgical decompression. Surgery is routinely planned if thrombolysis is successful in our practice for venous TOS. Thrombolysis was performed via

ipsilateral venous access with glidewire/glidecatheter crossing of the occlusion. Confirmation of placement of the catheter in the superior vena cava was routinely performed prior to placing a catheter for thrombolytic infusion. Images were obtained via the peripheral sheath, and via the catheter in the subclavian vein. Typically, Craig-MacNamera catheters were utilized for infusion of thrombolytic agents. The type of agent for thrombolysis varied over the course of the study. Patients were routinely brought back for reevaluation at 12–24 h after beginning infusion, and continued until complete lysis was achieved. Patients did not undergo stenting of the residual stenosis at the time of initial venography. Patients were routinely placed on anti-coagulation for 3–6 weeks prior to open surgical intervention, and counseled to avoid activities that would compress the subclavian vein prior to surgery. Patients were evaluated with venography at the time of surgery, prior to surgical resection of the rib, to confirm continued patency of the vein, and after resection to reevaluate for the need for angioplasty or repair of the vein. Open venorraphy or venous stenting was not required in any of the venorrhaphy patients. Hypercoagulable workup was not routinely performed on any of the patients. Several of the arterial patients who presented with acute ischemia of the arm underwent embolectomy via brachial artery cutdowns at a preceeding operation. Standard embolectomy was performed on these patients with subsequent evaluation for the source of emboli.

**Statistical analysis**

IRB approval was obtained prior to undertaking the study. Data was analysed using Microsoft Excel spreadsheet. Statistical evaluation included student’s t-test for independent samples. Significance was taken at the p < 0.05 level.

**Results**

We identified 12 patients with early-onset TOS in which 13 operations were performed over this 14 year period. One patient had been identified at 3 separate institutions as having bilateral neurogenic TOS, and bilateral cervical ribs. She successfully underwent surgery bilaterally in a staged process. The mean age at presentation was 16.8 ± 2.5 (12–20) (Table 1). The patient distribution was noted to be 66.67% female and 33.33% male. Patients were discharged on average 2.3 days after surgery. There was only one complication of a pneumothorax managed by tube thoracostomy with rapid resolution. Table 2 depicts the distribution of all TOS patients operated during the same time period.

The pattern of distribution of TOS symptoms in the first two decades was 38% arterial, 38% neurogenic and 24% venous. Time from onset of symptoms to treatment was 15.7 ± 21.6 months (2 days–60 months). Patients with neurogenic symptoms had a more significant delay: 34 ± 23.7 months (3–60 months) versus arterial: 2.2 ± 1.9 months (2 days–5 months) and venous: 7.6 ± 3.1 months (5–12months, p < .001).

Multiple diagnostic testing modalities were utilized for early-onset TOS. Patients with neurogenic TOS symptoms
had a mean of 2.6 test performed while patients with arterial or venous TOS had 2.25 ($p < 0.05$) test performed. Tests for neurogenic symptoms included cervical spine films, MRI, CT, EMG, and duplex while patients with venous symptoms underwent duplex and venography. All but 2 of the patients with NTOS had cervical ribs associated with their symptoms. Patients with arterial symptoms underwent duplex, CT, echocardiogram, and angiograms. Two of the patients with the arterial subtype underwent brachial embolectomy due to arm ischemia as their original presentation. Patients with alternate diagnoses by CT, MRI, EMG such as cervical compression or peripheral entrapment were not treated for TOS, and are not included in this series. Patients with arterial symptoms were evaluated for embolic sources, and none were found to have cardiac or other sources other than the compressed subclavian artery.

All patients were treated with outpatient postoperative rehabilitation for a mean of 5.3 ± 1.8 weeks (4–8 weeks). There was no statistical difference in the amount of rehabilitation required for neurogenic versus vascular TOS.

The average time to symptom resolution overall was 10.9 ± 8.9 weeks. Symptom resolution was determined by a complete improvement in both physical findings and symptomatic relief by subjective report. There was a significant time to symptom resolution by gender. Males required 2.25 ± 1.3 weeks while females required 14 ± 8.6 weeks ($p = .006$). The sample was evenly distributed for indication between groups.

The duration of symptoms prior to surgery had an impact on the time of symptom resolution after the procedure. Intervention within 3 months of onset of symptoms did not extend symptom resolution time ($p = .16$). However, if there was a delay of more than 6 months prior to surgery then the time to symptom resolution was lengthened ($p = .049$).

Two-thirds of patients responded to a follow-up questionnaire. The group that responded was comprised of 62% arterial TOS, 25% neurogenic TOS, and 12.5% venous TOS (Table 3). The mean time to survey from intervention was 26.6 ± 17.5 months (2–66 months). All respondents remained symptom free at last follow-up and by questionnaire. Narcotic pain medication was necessary for only 3.0 ± 2.3 weeks (1–8 weeks). No additional surgical interventions were necessary.

All of the patients that responded to the follow-up survey were students at the time of the operation. Twenty-five percent reported an increase in activity including sports as compared to their condition prior to intervention. In addition, 33.33% were able to participate in work activity that they were not able to perform prior to surgery (Table 4). Of the students who had neurogenic TOS, all had limited activity preoperatively due to their upper extremity symptoms. After intervention, all patients with neurogenic TOS who responded currently had jobs and were able to function at a higher level in athletic endeavors after the operation. The patients with venous and arterial TOS frequently did not have limited activity preoperatively due to the rapid onset of symptoms and intervention, and therefore, simply returned to normal activity without an increase from the pre-operative status.

### Table 1

<table>
<thead>
<tr>
<th>Age at onset</th>
<th>Type</th>
<th>Diagnosis delay</th>
<th>Simultaneous additional procedures</th>
</tr>
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<tbody>
<tr>
<td>20</td>
<td>Neuro</td>
<td>12 mo</td>
<td>Resection Cervical rib</td>
</tr>
<tr>
<td>19</td>
<td>Neuro</td>
<td>35 mo</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>Neuro</td>
<td>60 mo</td>
<td>Resection Cervical rib</td>
</tr>
<tr>
<td>13</td>
<td>Neuro</td>
<td>66 mo</td>
<td>Resection Cervical rib</td>
</tr>
<tr>
<td>12</td>
<td>Neuro</td>
<td>3 mo</td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>Arterial</td>
<td>1 mo</td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>Arterial</td>
<td>2 mo</td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>Arterial</td>
<td>5 mo</td>
<td>Resection subclavian aneurysm/bypass</td>
</tr>
<tr>
<td>17</td>
<td>Arterial</td>
<td>2 days</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>Arterial</td>
<td>3 mo</td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>Venous</td>
<td>12 mo</td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>Venous</td>
<td>5 mo</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>Venous</td>
<td>6 mo</td>
<td></td>
</tr>
</tbody>
</table>

### Table 2

**Age at surgical procedure for TOS (1994–2007).**

### Table 3

<table>
<thead>
<tr>
<th>TOS type</th>
<th>Response</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arterial</td>
<td>62%</td>
</tr>
<tr>
<td>Neurogenic</td>
<td>25%</td>
</tr>
<tr>
<td>Venous</td>
<td>12.5%</td>
</tr>
</tbody>
</table>

### Table 4

**Follow-up results.**

| Improvement or resolution of Symptoms | 100% |
| Increased ability to work | 33.3% |
| Increased athletics | 25% |

Discussion

Thoracic outlet syndrome 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. It is more commonly seen in women, and typically is reported to occur between the 3rd and 5th decades of life, despite the fact that many episodes are associated with anatomical abnormalities.
The gender bias was also seen in the early-onset population in our review. The anatomy of the thoracic outlet in a child is constantly changing with growth and development, as was first pointed out by Todd in 1912.7,8 The embryological development of this area is therefore important to appreciate and understand to explain the symptoms from thoracic outlet and their association with congenital anomalies.9–11

We were unable to recognize a clear pattern or method of diagnosis. Arterial and venous TOS are usually not difficult to recognize and the diagnosis can be confirmed by angiography. The diagnosis of neurogenic TOS is more challenging because its symptoms of nerve compression are not unique. The clinical diagnosis relies on documenting several positive findings on physical examination. To date there is still no reliable objective test to confirm the diagnosis, as noted by Saunders et al.12

We found early-onset TOS to have a significantly different distribution of disease than typical onset. Early-onset TOS had only a 38% incidence of neurogenic symptoms while typical onset ranged from 94 to 97%.11,13 Arterial TOS represented 38% and venous TOS represented 24% of the early-onset TOS patients, while vascular subtypes of TOS only occur in a very small percent of the typical TOS patients.11 This finding is supported by Arthur et al. who noted that 52% had vascular TOS and 44% had neurogenic TOS.14 We believe that there are more congenital abnormalities accounting for symptoms in the early-onset population with true impingement on vascular structures. Adults often present after minor repetitive trauma or major trauma with alterations to the muscle as the primary etiology of symptoms. This may account for the differing distribution of subtypes seen in the two populations.

TOS is often not readily recognized in adults due to the lack of familiarity of many primary care specialists, even when patients present with classic symptoms. The diagnosis of neurogenic TOS remains difficult as there is no single clinical or objective test that has been accepted as definitely establishing the diagnosis of this most common subtype seen in the typical onset population. Diagnosis in early-onset TOS may be even more difficult as TOS is not thought of as a disorder afflicting people in the first two decades of life, and pediatricians are even less likely to consider this disorder as the cause of patients’ symptoms. Young people are also more likely to ignore or downplay symptoms as being simple muscle strain or not express their symptoms as clearly as adults, thereby leading to delay in diagnosis. Once young patients presented to a physician, an average of 2.6 tests were performed before the diagnosis was made. Even in typical onset TOS, the diagnosis is rarely disputed for arterial or venous symptoms in which classic radiological findings are present. The area in which diagnosis is most complicated and disputed is in the arena of NTOS, for which there is no one diagnostic test, and issues of secondary gain are often considered.

Our general approach has been a supraclavicular approach for thoracic outlet decompression.15 Some recommend medial claviculectomy or transaxillary first rib resection,16 and others recommend a combined approach that employs a dual exposure.17 We feel that this approach allows for wide exposure, more access to anatomic structures in the thoracic outlet, and a more complete resection of a cervical rib.

We believe that early-onset TOS patients should be treated aggressively with early surgical intervention for arterial and venous types. This includes thrombolysis and embolectomy when indicated in addition to first rib resection, anterior scalenectomy, cervical rib resection when present, and excision of anomalous bands. This concurs with the findings of Melby et al. who evaluated outcomes of venous TOS in competitive athletes, with a median age of 20.3 years. Optimal outcomes were found to depend on early recognition by treating physicians and prompt referral for comprehensive surgical management.18 Vercellio et al. concurs that the same surgical strategy in adult patients is advisable for affected children.19 We still believe in a trial of physical therapy, anti-inflammatories and muscle relaxants for those with NTOS prior to surgical intervention. We have seen a small percent of early-onset NTOS patients respond to medical intervention and not require these patients respond and not require surgical intervention, however this has been uncommon in our experience. Young patients tolerate surgery well and on average they required minimal rehabilitation before returning to normal activities, and minimal narcotic pain medications. Of the respondents, no additional surgery or narcotic pain medication was needed after recovery from the initial TOS surgery. This finding is significantly different than studies in typical onset adults, who frequently have prolonged recoveries, and may relapse after initial improvement up to 2 years after surgery.20 Some of the hesitance to intervene surgically in NTOS stems from poor outcomes believed to be related to secondary gain issues. We believe that secondary gain issues are uncommon or even rare in the young patients, possibly explaining the excellent outcomes even of NTOS, with many being active athletes. The activity level of the young patients may also assist in more rapid recovery with patients typically being in better overall physiologic condition than their older counterparts. Our patients were able to return to function quickly. Resolution of symptoms was usually noted by two months. All patients felt the operation was beneficial. Early-onset patients were able to return to school, work or sports soon after the operation. Even patients with neurogenic TOS were able to participate more in gym and activities with their peers than prior surgery.

TOS should be considered in the diagnosis of patients in the first two decades of life who develop upper extremity symptoms, especially if they are involved in athletic endeavors which may precipitate its development. It should even be a consideration in patients with a history of trauma in the shoulder girdle.21 Delay of diagnosis is commonly noted, and it is possible that NTOS is underdiagnosed in this population. Vascular TOS is more commonly identified in young people than in typical onset TOS, and is actually more frequent than neurogenic TOS in this population. Unlike with adults, early-onset patients have excellent long-term results from surgical intervention, with complete and lasting symptom resolution. Surgery for TOS should be considered more rapidly
in early-onset patients to allow return to normal function.

**Conflict of Interest/Funding**

None.

**References**

7. Todd TW. Cervical rib factors controlling its presence and its size, its bearing on the morphology of the shoulder, with four cases. *J Anat Physiol* 1911–1912;45:293–304.