Long-term outcomes after surgical treatment of pediatric neurogenic thoracic outlet syndrome

Jennifer Hong, MD,¹ Jared M. Pisapia, MD,² Zarina S. Ali, MD,² Austin J. Heuer,³ Erin Alexander, BS,³ Gregory G. Heuer, MD, PhD,^{2,3} and Eric L. Zager, MD²

¹Division of Neurosurgery, Dartmouth-Hitchcock Medical Center, Lebanon, New Hampshire; ²Department of Neurosurgery, University of Pennsylvania, Philadelphia; and ³Division of Neurosurgery, The Children's Hospital of Philadelphia, Pennsylvania

OBJECTIVE Neurogenic thoracic outlet syndrome (nTOS) is an uncommon compression syndrome of the brachial plexus that presents with pain, sensory changes, and motor weakness in the affected limb. The authors reviewed the clinical presentations and outcomes in their series of pediatric patients with surgically treated nTOS over a 6-year period.

METHODS Cases of nTOS in patients age 18 years or younger were extracted for analysis from a prospective database of peripheral nerve operations. Baseline patient characteristics, imaging and neurophysiological data, operative findings, and outcomes and complications were assessed.

RESULTS Twelve patients with 14 cases of nTOS surgically treated between April 2010 and December 2016 were identified. One-third of the patients were male, and 2 male patients underwent staged, bilateral procedures. Disabling pain (both local and radiating) was the most common presenting symptom (100%), followed by numbness (35.7%), then tingling (28.6%). The mean duration of symptoms prior to surgery was 15.8 ± 6.6 months (mean $\pm$ SD). Sports-related onset of symptoms was seen in 78.6% of cases. Imaging revealed cervical ribs in 4 cases, prominent C-7 transverse processes in 4 cases, abnormal first thoracic ribs in 2 cases, and absence of bony anomalies in 4 cases. Neurophysiological testing results were normal in 85.7% of cases. Conservative management failed in all patients, with 5 patients reporting minimal improvement in symptoms with physical therapy. With a mean follow-up after surgery of 22 ± 18.3 months (mean $\pm$ SD), pain relief was excellent ($> 90\%$) in 8 cases (57.1%), and good (improved $> 50\%$) in 6 cases (42.9%). On univariate analysis, patients who reported excellent pain resolution following surgery at long-term follow-up were found to be significantly younger, and to have suffered a shorter duration of preoperative symptoms than patients who had worse outcomes. Lack of significant trauma or previous surgery to the affected arm was also associated with excellent outcomes. There were 4 minor complications in 3 patients within 30 days of surgery: 1 patient developed a small pneumothorax that resolved spontaneously; 1 patient suffered a transient increase in pain requiring consultation, followed by hiccups for a period of 3 hours that resolved spontaneously; and 1 patient fell at home, with transient increased pain in the surgically treated extremity. There were no new neurological deficits, wound infections, deep vein thromboses, or readmissions.

CONCLUSION Pediatric nTOS commonly presents with disabling pain and is more frequently associated with bony anomalies compared with adult nTOS. In carefully selected patients, surgical decompression of the brachial plexus results in excellent pain relief, which is more likely to be seen in younger patients who present for early surgical evaluation.

<https://thejns.org/doi/abs/10.3171/2017.7.PEDS17257>

KEY WORDS thoracic outlet syndrome; brachial plexus; pediatric; anterior scalenectomy; peripheral nerve; pain

THORACIC outlet syndrome (TOS) is a neurovascular compression syndrome caused by constriction of the brachial plexus or subclavian artery and vein as they exit the thoracic outlet. This syndrome can have a variable presentation, and symptoms are typically divided into vascular or neurogenic, although an individual may have a mixture of both. Neurogenic TOS (nTOS) has a

wide constellation of symptoms including pain, paresthesias, and motor deficits in the neck and/or extremity.²⁶ The diagnosis of TOS is controversial, because the definition describes the location of the compression but not a specific pathogenesis.

In adults, the incidence of TOS is reported to be 0.3%–2% of the general population between the ages of 25 and

ABBREVIATIONS EMG = electromyography; NCS = nerve conduction studies; nTOS = neurogenic thoracic outlet syndrome; POD = postoperative day; TP = transverse process; UEFI = upper-extremity functional index; US = ultrasound.

SUBMITTED May 15, 2017. **ACCEPTED** July 21, 2017.

INCLUDE WHEN CITING Published online November 10, 2017; DOI: 10.3171/2017.7.PEDS17257.

40 years, with > 80% of patients falling between those ages.¹⁸ Women are 3 times more likely to be affected than men. Thoracic outlet syndrome also affects children, and appears to be much rarer in this age group, although the true incidence is unknown. Pediatric or early-onset TOS is poorly understood; the literature to date consists of small case series with limited follow-up.^{2,11,13,14,16,21–23,26–30} There may be several reasons for this paucity, including a lack of familiarity with TOS among pediatricians, minimization of symptoms by children, or difficulty communicating symptoms to adults in young children. Nevertheless, TOS in children and young adults remains understudied.

Management of pediatric TOS is therefore challenging, with no clear guidelines for selecting patients for intervention. Unlike adult TOS, which is overwhelmingly neurogenic (95%) in nature,^{8,19} pediatric TOS is often vascular, with only one-third of patients presenting with neurogenic symptoms.²⁹ Compared with adult nTOS, pediatric nTOS is less prevalent, more difficult to diagnose, and more likely to have a distinct pathogenesis and natural history. Whether the same treatment strategies used in adult TOS are effective in children is debatable, particularly in young children who are still developing. Data on outcomes after treatment of TOS in children are therefore important to analyze, especially in patients with long-term follow-up.

We undertook a retrospective review of children presenting to 2 institutions. Particular attention was paid to the role of conservative treatment, intraoperative findings, and clinical follow-up.

Methods

Participants and Study Design

Following approval by the institutional review boards of the University of Pennsylvania and Children's Hospital of Philadelphia, hospital records and a prospectively maintained database of all peripheral nerve operations at these 2 institutions were queried for TOS and ICD-9 codes 21700 and 64713 between April 2010 and July 2016. A diagnosis of TOS was made based on hospital discharge documentation and referral information.

Clinical Variables and Outcomes Assessment

Clinical data were retrieved through retrospective chart review. Data available included age, sex, presenting symptoms, clinical examination results, imaging findings, electrophysiology studies, operative findings, and clinical follow-up.

Work-Up and Management

Children referred to the Hospital of the University of Pennsylvania or Children's Hospital of Philadelphia for evaluation of possible TOS underwent a complete history and a detailed physical and neurological examination. Plain chest or cervical spine radiographs were obtained in all patients to rule out a cervical rib (Fig. 1). Electromyography (EMG) and nerve conduction studies (NCS) were obtained in all patients. All patients except for 2 who underwent MRI of the cervical spine or affected brachial plexus. A chest CT was obtained in 2 patients to better evaluate bony anomalies. Ultrasound (US) of the affected

FIG. 1. Plain chest radiograph (A) and reconstructed chest CT (B) demonstrating a right cervical rib, with fusion to the first thoracic rib and resulting pseudarthrosis in a 12-year-old girl with right nTOS.

brachial plexus with dynamic maneuvers with or without US-guided steroid injection of the brachial plexus was obtained when available and if the patient and parents were amenable. If vascular symptoms were present, US of the vessels of the thoracic outlet was obtained. All patients underwent a trial of physical therapy for at least 4 weeks (many for 6 months) prior to surgical intervention.

Surgical Approach and Follow-Up

All patients underwent an anterior supraclavicular brachial plexus exploration, with anterior scalenectomy and, when present, resection of cervical rib or other bony sites of compression (Fig. 2). Neuromonitoring with a handheld stimulator as well as traditional console-based monitoring (compound muscle action potentials and somatosensory evoked potentials) was used in all cases. The patients were treated as previously described.⁹

All patients were discharged home once discharge criteria were met, typically on postoperative day (POD) 1. All patients returned to clinic for routine follow-up at 4–6 weeks, or as travel schedules allowed. No routine repeat imaging was obtained.

Long-term telephone follow-up was attempted for all patients. A standardized questionnaire asking patients to rate their pain relief, residual numbness or tingling, motor weakness, and satisfaction with the operation was administered. The upper-extremity functional index (UEFI)^{7,24} was also administered to all patients, and they were asked whether they would recommend surgery to other patients with nTOS. Three attempts were made on separate dates to contact patients. When the patient was not available to answer questions, follow-up data were obtained from a parent.

Statistical Analysis

Statistical analysis was conducted with commercially available R software (www.r-project.org). Contingency tables were created and chi-square values were calculated. The mean is expressed $\pm$ SD throughout.

Univariate analyses were performed with the Student t-test (2-tailed) for numeric variables and with the Fisher exact test (2-tailed) for categorical variables to determine significant associations between these variables and an excellent outcome, defined as >90% relief of painful symptoms following surgery. A p value of ≤ 0.05 was consid-

FIG. 2. Intraoperative views of supraclavicular brachial plexus exploration, anterior scalenectomy, and resection of the cervical rib and pseudarthrosis in the same patient. **A:** The black arrowhead indicates the pseudarthrosis at the junction of the cervical rib and first rib. The white arrow indicates the cut edge of the anterior scalene. The subclavian artery is displaced superiorly by the cervical rib. The bottom vessel loop is placed around the lower trunk of the brachial plexus, which is displaced inferiorly. **B:** Photograph obtained after resection of the cervical rib and associated pseudarthrosis. The white arrow indicates the cut edge of the anterior scalene. The subclavian artery has returned to its normal course. Bottom vessel loop is placed around the lower trunk of the brachial plexus, which has also returned to its normal course. S = subclavian artery.

ered statistically significant. Multivariate analysis was not pursued due to the small size of the population.

Results

Population Demographics and Presentation

Fourteen cases of nTOS were reviewed in 12 children (Table 1). The mean age at time of surgery was 15.2 ± 2.8 years, with an age range from 8 to 18 years. One-third of the patients were male, with 2 male patients undergoing staged, bilateral procedures (Cases 1 and 3).

Presenting symptoms were primarily pain (in all patients) of the neck, shoulder, arm, and hand. Patients also presented with numbness (35.7% of cases), and tingling (28.6% of cases). Three patients (21.4% of cases) had concurrent vascular symptoms including mottling or swelling of the arm and hand. Half of all cases involved the dominant hand, with 2 patients suffering bilateral presentations that were separated by time periods of 9 months in one, and 4 years in the other. The mean duration of symptoms prior to treatment was 15.8 ± 6.6 months.

A history of significant participation in sports, in which playing sports was associated with exacerbation or onset of nTOS symptoms, was seen in 78.6% of cases—with 71.4% of cases occurring in sports that involve the upper extremity (i.e., swimming, baseball, crew, water polo). Trauma was associated with 28.6% of cases and included motor vehicle accident, sports injury, and previous surgery.

On examination, 71.4% of the cases of pediatric nTOS exhibited a positive Roos maneuver and 64.3% demonstrated a positive Tinel's sign. Patients reported tenderness to palpation over the brachial plexus in 57.1% of cases and numbness in the hand in 28.6% of cases. Twelve cases (85.7%) had at least one positive examination finding. Although some patients complained of subjective weakness, all patients were at full strength to confrontation and demonstrated normal reflexes on examination.

Imaging Findings and Electrophysiology

All patients underwent chest or cervical spine radiographs to evaluate for the presence of cervical ribs or prominent transverse processes (TPs). In our 14 cases, we observed 4 cervical ribs (28.6%), 4 prominent TPs of the lower cervical vertebra (28.6%), and 2 first rib anomalies, namely a fused first and second rib and a partially resected first rib (14.3%). Four cases did not have any bony anomalies. Additional imaging included MRI of the cervical spine in 8 patients, which demonstrated a congenital fusion of C-2 and C-3 in 1 patient, and a mild C5–6 disc bulge in another; MRI of the brachial plexus was performed in 6 patients, demonstrating an intramuscular course of the brachial plexus in 1 patient (through the anterior scalene muscle) and postoperative changes from previous transaxillary rib resection in another patient. The CT scans of the chest in 2 patients demonstrated bilateral cervical ribs in one patient and a synchondrosis of the ipsilateral first and second ribs in another.

Overall, the yield of MRI of the cervical spine and brachial plexus for abnormalities was 25%, although it is unclear how much these abnormalities contributed to patient symptomatology. Ultrasound of the brachial plexus with dynamic maneuvers and possible steroid injection of the plexus was obtained in 9 cases (64.3%). This demonstrated 2 cases with dynamic neural compression (i.e., when the arm was elevated), 2 cases of dynamic arterial compression, and 1 diagnosis of bilateral vascular TOS by wrist indices. Ultrasound was the most sensitive imaging study for compression, with positive findings of concern for compression in 50% of patients. Of 4 patients who underwent US-guided steroid injections, 3 (75%) reported transient reduction in pain, and 1 noticed no changes.

All patients underwent EMG (Table 2) and NCS. Two studies (14.2%) were abnormal, with one study demonstrating a mild plexopathy of the lower trunk, and another with mild bilateral C-5 radiculopathy.

TABLE 1. Demographic data and clinical presentation in 14 cases of nTOS

Case No.*	Age (yrs)	Sex	Preop		Presenting Complaint	Vascular Symptoms	Sports History	Past Medical History	Provocative Maneuvers	Exam Findings
			Duration of Symptoms (mos)	Dominant Arm Affected?						
1a	17	M	24	Yes	Pain	No	Crew	Concussion	+ Roos, + TTP	Full strength, normal reflexes, numbness of 4th & 5th digits
1b	17		15	No	Pain	No	Crew	Concussion	+ Roos, + TTP	Full strength, normal reflexes, TTP over supraclavicular fossa
2	16	F	11	Yes	Pain, numbness, weakness	No	None	Asthma	+ Roos, + Tinel's	Full strength, normal reflexes, numbness of hand
3a	8	M	19	Yes	Pain, numbness	No	Baseball	Asthma	ND	Full strength, normal reflexes
3b	12		9	No	Pain, numbness	No	Baseball	Asthma	ND	Full strength, normal reflexes
4	16	F	24	Yes	Pain, numbness	No	Softball	Polycystic ovarian syndrome	+ Roos	Full strength, normal reflexes, numbness of 1st, 2nd, & 3rd digits
5	15	F	13	No	Pain	No	None	Prior shoulder surgery	+ Tinel's, + TTP	Full strength, normal reflexes
6	17	M	18	Yes	Pain, tingling	Mottling of hands	Hockey	None	+ Roos, + Tinel's, + TTP	Full strength, normal reflexes, TTP over infraclavicular fossa
7	18	M	24	No	Pain, tingling	Swelling of arm	Baseball	Sternum & clavicle fracture, prior TARR	+ Roos, + Tinel's, + TTP	Full strength, normal reflexes, TTP over infraclavicular fossa
8	17	F	7	No	Pain, numbness	No	None	MVA	+ Roos, + Tinel's, + TTP	Full strength, normal reflexes, TTP over infraclavicular fossa
9	12	F	24	Yes	Pain, tingling	No	Swimming	Asthma	+ Tinel's, + TTP	Full strength, normal reflexes, TTP over supraclavicular fossa
10	15	F	6	No	Pain, tingling	Arm discoloration	Lacrosse, water polo	ADHD	+ Roos, + Tinel's	Full strength, normal reflexes
11	15	F	11	Yes	Pain	No	Soccer	None	+ Roos, + Tinel's, + TTP	Full strength, normal reflexes
12	18	F	16	No	Pain	No	Field hockey	Sports injury	+ Roos, + Tinel's	Full strength, normal reflexes, numbness of 4th & 5th digits
Mean ± SD	15.2 ± 2.8		15.8 ± 6.6							
Range	8–18		6–24							
Summary	66.6% F		50% dominant arm			21.4% vascular symptoms	78.6% sports history		71.4% + Roos, 64.3% + Tinel's, 57.1% + TTP	

ADHD = attention deficit hyperactivity disorder; Exam = examination; MVA = motor vehicle accident; ND = not done; TARR = transaxillary first rib resection; TTP = tenderness to palpation; + = positive.

* Case 1 and 3 patients underwent staged, bilateral procedures.

TABLE 2. Imaging and electrophysiology in pediatric nTOS

Case No.*	MRI of Cervical Spine	MRI of Brachial Plexus	CT of Chest	Radiography	US	EMG	Bony Anomalies on Affected Side
1a	None	Normal, prominent musculature	None	Bilat prominent C-7 TP	Normal	Normal	Elongated C-7 TP
1b	None	Normal, prominent musculature	None	NP	Normal	Normal	Elongated C-7 TP
2	Bilat cervical ribs at C-7 & C-6, w/ fusion of multiple ribs on rt	None	None	Right C-6 riblet & partial C-7 rib, complete lt C-7 rib, partial fusion of rt T-1 & T-2 ribs	Apposition of the brachial plexus & the cervical rib on the rt, w/ dynamic movement	Mild plexopathy of lt lower trunk	Cervical rib
3a	Normal	Normal	None	Bilat cervical ribs, rt > lt	None	Normal	Cervical rib
3b	NP	Normal	None	NP	None	Normal	Cervical rib
4	Normal	None	None	Bilat slight enlargement of C-7 TP	Normal	Normal	Elongated C-7 TP
5	None	None	None	Normal	Normal	Normal	None
6	Congenital fusion of C2–3	Intramuscular course of C-5 & C-6, bilat enlargement of subclavius muscles	None	C2–3 fusion	Bilat compression of the brachial plexus w/ dynamic movement	Bilat mild C-5 radiculopathy	None
7	None	Expected postop changes, elongated C-7 TP, lt > rt, not contacting the middle trunk, normal plexus	None	Status postresection for lt 1st rib	Normal	Normal	Remnant of lt 1st rib
8	Normal	None	Synchondrosis of lt 1st & 2nd rib	Normal	Wrist indices demonstrate bilat TOS	Normal	Synchondrosis of lt 1st & 2nd ribs
9	None	None	Bilat cervical ribs, rt much larger than lt; rt rib forming pseudarthrosis, w/ thickened, expanded T-1 rib	Bilat cervical ribs, rt > lt	None	Normal	Cervical rib w/ pseudarthrosis
10	Normal	Normal	None	Normal	Compression of subclavian artery & vein w/ dynamic movement	Normal	None
11	Normal	None	None	Normal	None	Normal	None
12	Mild C5–6 disc bulge	Normal	None	Lt C-7 TP slightly enlarged	Mild enlargement of roots, trunks, & divisions of lt brachial plexus, subclavian artery btwn middle & lower trunk, compression of vascular structures w/ dynamic movement	Normal	Prominent lt C-7 TP

NP = not performed.

* Case 1 and 3 patients underwent staged, bilateral procedures.

Neurosurgical Interventions and Outcomes

All patients underwent anterior supraclavicular brachial plexus exploration and anterior scalenectomy (Table 3). Furthermore, additional compressive soft tissue and bony

structures were removed if they were noted to be abutting nerves at the time of surgery. This included resection of a rib or prominent TP in 5 cases (35.7%), division of Sibson's fascia (suprapleural membrane) in 3 cases (21.4%), and a

TABLE 3. Outcomes following surgical intervention for pediatric nTOS

Case No.*	Duration of FU (mos)	Intraop Findings	LOS (days)	Pain Relief (%)	Numbness, Tingling	UEFI Score	Surgery Rec	30-Day Complications
1a	12.5	Fibrous bands in the anterior scalene	2	70	Occasional, improved	73	Yes	Transiently increased pain immediately postop requiring pain consult, hiccups for 3 hrs that resolved spontaneously on POD 3
1b	26	Fibrous bands in the anterior scalene	1	85	Occasional, improved	73	Yes	None
2	21	Hypertrophic anterior scalene muscle w/ fibrous firm bands	1	99	Occasional, w/ heavy activity	77	Yes	None
3a	59	Fibrous bands in the anterior scalene muscle, cervical rib	1	100	None	79	Yes	None
3b	21	Hypertrophic anterior scalene muscle, compressive Sibson's fascia, tight fascial band btwn cervical rib & middle scalene muscle	1	100	None	79	Yes	Pneumothorax observed at OSH on POD 4
4	28	Fibrous bands in the anterior scalene muscle, Sibson's fascia compressing lower trunk	1	80	Occasional w/ heavy activity	65	Yes	Fall down stairs on POD 2
5	29	Fibrous bands in middle scalene muscle, compressive Sibson's fascia	2	80	None	68	Yes	None
6	42	Fibrous bands in the anterior scalene muscle	2	100	Occasional, improved	79	Yes	None
7	50	Inferior portion of anterior scalene muscle scarred & adherent to lower trunks, remnant of 1st rib	1	80	Occasional, improved	70	Yes	None
8	1	Displacement of brachial plexus by aberrant first thoracic rib	2	100	None	NA	NA	None
9	1	Displacement of brachial plexus by cervical rib	2	100	None	NA	NA	None
10	10	Fibrous bands in the anterior scalene muscle, compression of the middle & lower trunk	1	90	None	78	Yes	None
11	5	Hypertrophic anterior scalene muscle w/ fibrous bands, compression of lower & upper trunk by vascular structures	1	100	None	77	Yes	None
12	5	Elongated C-7 TP; fibrous band deep to the lower & middle trunks & extending from the TP to 1st rib involving the medial border of the middle scalene muscle, distorting the plexus	1	50	Occasional, improved	44	Yes	None
Mean $\pm$ SD	22.17 $\pm$ 18.29			88.14 $\pm$ 15.08		74.36 $\pm$ 9.94		

FU = follow-up; LOS = length of hospital stay; NA = not available; OSH = outside hospital; POD = postoperative day; Rec = recommended.

* Case 1 and 3 patients underwent staged, bilateral procedures.

middle scalenectomy in 1 case (7.14%). The most common observations intraoperatively included the presence of fibrous bands within the anterior scalene in 11 cases (78.6%); hypertrophied scalene muscles in 3 cases (21.4%); and frank distortion of the brachial plexus by adjacent soft tissue, vascular, or bony structures in 5 cases (35.7%). The patient with prior transaxillary first rib resection underwent external neurolysis of plexus elements with resection of the scar tissue that was thought to be related to his pain.

All patients were discharged to home following surgery, with approximately two-thirds of patients leaving on POD 1. There were 4 minor complications in 3 patients (21.4% of cases) within 30 days of surgery. One patient suffered a transient increase in pain following surgery requiring a consultation visit by the pain management service, and then developed hiccups for a period of 3 hours that spontaneously resolved; 1 patient was diagnosed with a small pneumothorax on POD 4 at another hospital and was conservatively managed; 1 patient fell at home on POD 2, with resulting increased pain in her surgically treated arm, which had resolved at long-term follow-up.

The mean follow-up duration for all patients was 22.17 ± 18.29 months, with a range of 1–59 months). Clinical and long-term telephone follow-up was available for 12 cases; clinical follow-up alone was available in 2 cases. At the last postsurgical follow-up, complete resolution of pain was reported in 6 cases (42.9%), > 90% reduction in pain was reported in 2 cases (14.3%), > 50% reduction in pain was reported in 5 cases (35.7%), and 50% reduction in pain was reported in 1 case (7.14%). The mean reduction in pain was $88.14\% \pm 15.08\%$. Tingling and numbness resolved in 7 cases (50%), decreased in 5 cases (35.7%), and was only present with heavy activity in 2 cases (14.3%). No patients reported worsening weakness following surgery.

Twelve cases were assessed using the UEFI at long-term telephone follow-up (Table 3). In 8 (72.7%) of these cases, patients scored 71 points or higher, which is equivalent to normal function. Four patients had clinically detectable decreased scores in the UEFI (< 71 points); however, 1 of these patients described worsened function after a fall at home on POD 2 following a period of initial improvement, and 3 of these patients had a history of injuries or surgery to the affected shoulder and arm, and denied that their upper-extremity function had changed noticeably post-operatively. All patients surveyed responded “yes” to the question of whether they would recommend surgery for other patients with nTOS.

Univariate Analysis

Univariate analysis was undertaken to identify predictors of an excellent outcome, defined as a pain reduction of $\geq 90\%$ (8 cases). With respect to continuous variables, patients who obtained $\geq 90\%$ pain relief were significantly younger than patients who did not, with a mean age of 14 ± 2.91 years versus 16.8 ± 1.17 years ($p = 0.04$). In addition, there was a trend toward a shorter duration of preoperative symptoms in the excellent outcomes group ($p = 0.08$) compared with the good outcomes group. With respect to categorical variables, absence of a prior injury or trauma to the affected arm (such as a sports injury, fracture, or prior surgery) was significantly associated with an excel-

lent outcome ($p = 0.05$). The presence of an abnormal rib, cervical or thoracic, showed a trend toward predicting excellent outcomes ($p = 0.13$), but neither this nor a history of sports participation were significantly associated with excellent outcomes (Table 4).

Discussion

Thoracic outlet syndrome is an uncommon clinical syndrome of pain and functional impairment resulting from compression of vascular and/or nervous structures, most often between the anterior and middle scalene muscles and the first rib. Neurogenic TOS, which often presents without clear examination, imaging, or electrophysiological findings, can be a diagnosis of exclusion. Thus, nTOS is difficult to study systematically, because no consensus diagnostic criteria exist, and the symptoms of nTOS frequently overlap with other diagnoses. Whereas nTOS in adults accounts for 95% of all TOS diagnoses, nTOS is only observed in 26% of pediatric TOS cases,^{2,18} resulting in an even smaller pool of data. The total number of reported pediatric nTOS cases in the literature to date is fewer than 100 (Table 5).^{2–5,11–13,16,20,21,25,26,28–30} Further complicating our understanding of nTOS in children is the unfamiliarity of many pediatricians with this clinical entity, the difficulty of younger children in trying to communicate neurological symptoms, the reluctance to pursue aggressive or invasive testing in children, and the uncertain natural history of nTOS in a population that undergoes rapid growth and dramatic hormonal changes.

We undertook a 6-year retrospective review of outcomes in children with nTOS following surgical decompression to assess the role for surgery and to identify predictors for excellent outcomes following surgery. Because standardized long-term outcomes are underreported in this patient population, we obtained long-term follow-up data on our patients by using a patient-reported functional

TABLE 4. Univariate analysis of factors associated with $\geq 90\%$ pain resolution following supraclavicular brachial plexus exploration and decompression

Variable	Excellent Outcome, n = 8	Good Outcome, n = 6	p Value
			Student t-test, 2-tailed
Age in yrs, mean $\pm$ SD	14 $\pm$ 2.9	16.8 $\pm$ 1.2	0.04*
Duration of symptoms prior to surgery in mos, mean $\pm$ SD	13.1 $\pm$ 6.44	19.3 $\pm$ 5.2	0.08
			Fisher exact test, 2-tailed
Sports history	7 (87.5)	5 (83.3)	0.52
Presence of abnormal rib	5 (62.5)	1 (16.7)	0.13
No previous surgery or trauma	8 (100)	3 (50)	0.05*

Unless otherwise indicated, values are expressed as the number of cases (%).

* Statistically significant.

TABLE 5. Literature review of outcomes following surgical intervention for pediatric nTOS

Authors & Year	No. of Pts	No. of Ops	No. of Cervical Ribs	Ages (yrs)	% Female	% w/ Sports History	Surgical Approach	% w/ Bony Anomaly	Mos of Follow-Up	% w/ Complete Response	Complications
Arthur et al., 2008	12	12	4	12–18	82	18	TARR	30	NR	40	1 reop for recurrent pain
Cağli et al., 2006	1	1	1	13	100	NR	SCBPD	100	1	100	None
Chang et al., 2011	9	9	1	13–18	88.9	77.8	TARR	11.10	15 (mean)	100	1 pneumothorax
Dahlin et al., 2009	2	2	2	11 & 16	100	50	SCBPD	100	12	100	None
Khan et al., 2012	1	1	1	13	100	NR	SCBPD	100	NR	100	None
Martins & Siqueira, 2007	1	1	1	9	100	100	SCBPD	100	NR	100	None
Maru et al., 2009	5	13	8	12–20	66.7	NR	SCBPD	60	2.5	100	1 pneumothorax
Rehemutula et al., 2015	10	7	2	4–13	76.9	NR	SCBPD	30	6–30	60	NR
Sanders et al., 2013	16	20	7	11–19	70	43	SCBPD	35	12–48	70	2 transient phrenic nerve palsies
Sen et al., 2007	1	1	1	8	0	NR	TARR	100	60	100	None
Swierczyńska et al., 2005	3	1	NR	12–14	100	NR	NR	NR	12	30	None
Teddy et al., 2012	13	20	14	9–14	76.9	100	17 SCBPD, 3 TARR	72	6–72	53.8	2 reops for persistent pain
Vercellio et al., 2003	2	2	2	12 & 16	50	50	SCBPD	100	24	100	None
Vu et al., 2014	4	4	4	4 mos–10 yrs	75	NR	SCBPD	100	3–36	100	None
Yang & Letts, 1996	4	3	3	12–13	NR	NR	TARR	75	24	100	None
Total	84	97	51								
Mean ± SD					77.6 ± 27.2%	62.6 ± 33.26%		74.2 ± 32.7%		83.5 ± 25.5%	

NR = not reported; Pts = patients; SCBPD = supraclavicular brachial plexus decompression, anterior scalenectomy, and resection of cervical rib when present.

index. We also attempted to better define the presenting characteristics and assess the utility of various diagnostic tests to assess for nTOS.

Pediatric nTOS is a Challenging Diagnosis

In our series, the average time to surgery from the onset of nTOS symptoms was > 1 year (15.8 ± 6.6 months), which is consistent with other series.^{4,16,26} This is partially due to the trial of physical therapy that our patients undergo prior to surgery, but it also reflects a delay in diagnosis due to extensive work-ups, consideration of other conditions, or failed pain management. A delay in surgical evaluation and management is understandable because initial imaging of the cervical spine and brachial plexus is frequently negative (approximately 75% in our series), and other diagnoses may be pursued in the absence of evidence

for injury or compression. The rate of negative MRI findings is also high in other similarly sized case series,^{2,26} and Sanders et al. state that they found MRI to be expensive and unhelpful, and that they no longer routinely order the test.²⁰ In our experience, US of the brachial plexus with dynamic maneuvers was more helpful in demonstrating compression of nervous and vascular structures (50% of cases), which might suggest an underlying mechanism for injury. With regard to the diagnostic utility and predictive power of a response to a US-guided steroid injection, our results were mixed (1 patient responded to injection and had an excellent outcome of surgery, 2 patients responded and had good outcomes, and 1 patient did not respond and had an excellent outcome).

Electrophysiological studies (i.e., EMG and NCS) were generally unhelpful in diagnosing nTOS in our cohort. This has also been observed in other series, where rates

of abnormal EMG and NCS findings range from 7.69% to 11% in children with nTOS.^{2-5,11-13,16,20,21,25,26,28-30} The reliability of provocative maneuvers is notoriously low, and our patients demonstrated a 71.4% likelihood of a positive Roos sign, a 64.3% likelihood of a positive Tinel's sign, and a 51.7% likelihood of tenderness to palpation over the brachial plexus. Although the majority of our patients exhibited 1 of these 3 findings on examination (12/14 cases, 85.7%), the presence of a positive provocative maneuver was not associated with a good surgical outcome. We did not report the percentage of positive findings with Adson's maneuvers because the Adson's test has been noted to be positive in 50% of the normal population.²

The most convincing evidence for nTOS in our experience is a combination of a clinical history of persistent debilitating pain, accompanied by numbness or paresthesias that are exacerbated by overhead arm movements, with a significant sports history and a plain cervical radiograph demonstrating a bony anomaly. These 4 factors were observed in 8 cases (57.1%), and 2 of these 4 factors were observed in all cases.

Surgical Treatment is Effective and Safe for Pediatric nTOS

In our cohort, 57.1% of our cases (8 patients) reported > 90% reduction in pain at a mean follow-up period of 22 months after surgical decompression of the thoracic outlet. If patients who reported > 50% pain relief are also considered responders to surgery, the success rate of surgical decompression in our series increases to 92%. This high rate of durable pain relief is consistent with what has previously been observed in case series with > 5 cases of pediatric nTOS (mean complete response rate of 63.3%).^{2,4,13,16,20} Furthermore, there were no major 30-day complications in our patients. In the literature, the most commonly observed complications are transient phrenic nerve injuries (2%) and pneumothoraces (2%). Whereas a 3% reoperation rate was seen in previous studies (Table 5), no patients required reoperation in our group for reexploration of the brachial plexus during the follow-up period. However, it is possible that patients with short-term follow-up may have yet to present with recurrent symptoms, because the average time to recurrence was 18 months in a large adult series.¹ Two patients did elect to undergo subsequent orthopedic shoulder operations on the same side as their nTOS. Although both of these patients failed to obtain > 90% pain relief after nTOS surgery, they did recommend the surgery to other patients at long-term follow-up. This highlights the challenge of treating nTOS symptoms, which rarely respond to conservative management. Even partial pain relief with surgery results in high patient satisfaction, although frank counseling about the anticipated outcomes and risks of surgery is essential.

The role and extent of bone resection in surgical treatment for nTOS is debated. One series reported that more complications were associated with bone resection,²⁶ leading some surgeons to advocate for soft-tissue resection alone, even in cases with cervical ribs.¹⁷ We prefer the supraclavicular approach in part because it provides excellent outcomes in many patients without bone removal; however, if the brachial plexus is deviated by bone after

anterior scalenectomy, bone removal may be required to adequately decompress the neural elements. We do not recommend routine removal of any abnormal bony element. For example, enlarged C-7 TPs are not resected unless they contact the brachial plexus.

Of the patients who did not attain excellent pain relief following surgery, the reasons for failure are unclear. Some groups have invoked the double-crush hypothesis to explain persistent symptoms,²⁰ suggesting that a brachial plexus exploration alone may not address other superimposed sites of entrapment. However, only 1 of our unresponsive patients had an abnormal result on EMG that was consistent with C-5 radiculopathy. No other EMG or examination findings demonstrated additional sites of entrapment in the 5 other cases with residual pain after surgery. More likely, diagnostic uncertainty in combination with potential reinjury (1 of our patients fell on her surgically treated extremity shortly after discharge from the hospital, and 2 other patients had additional orthopedic shoulder surgeries) is at fault. In our experience, patients with < 90% pain relief after surgery at long-term follow-up were significantly more likely to have a history of trauma or prior surgery to the affected limb, which could result in complex, painful injuries to neighboring structures or the development of chronic pain syndromes in addition to damage to the brachial plexus. Based on this group, we now counsel pediatric patients with nTOS who have had prior trauma or surgery that brachial plexus decompression alone is less likely to result in an excellent outcome. It is also possible that some patients had compression or irritation of the plexus elements outside of the interscalene triangle. We did not routinely explore the infraclavicular plexus, because this is an extremely rare site of TOS. We have not pursued additional evaluation for these patients because they report very high satisfaction with surgery, despite less than excellent pain outcomes.

Careful selection of patients who will most likely benefit from surgery is critical. In our analysis of factors associated with complete resolution of pain after surgery, the presence of abnormal ribs on imaging trended toward statistical significance, an association that may have been more apparent with larger numbers of patients. This suggests that an anatomical predisposition to entrapment is a risk for development of pediatric nTOS, and surgical decompression has a greater likelihood of success when patients have static compression in addition to dynamic compression.

Unfortunately, there is an absence of good observational data about long-term results of conservative management as compared with surgery. Only 2 papers report positive outcomes for children who underwent conservative management. Rehemutula et al. described 3 children (ages 6, 6, and 7 years) who presented with nTOS and underwent intensive physical therapy and posture correction with local analgesic blocks of the anterior and middle scalene. At a minimum of 6 months of follow-up, these children were described as having satisfactory outcomes.¹⁶ Yang and Letts elected to observe 2 children with nTOS, ages 12 and 13 years, who did not have cervical ribs on imaging. Over a period of 2 years, they reported that the pain symptoms resolved with avoidance of the inciting arm po-

sitions.³⁰ In each case, the children whose nTOS was managed successfully with conservative measures were earlier-presenting patients with relatively mild symptoms. In light of these findings, we recommend a trial of conservative management in all patients prior to consideration for surgery. A few of our patients undergo US-guided steroid injections of the brachial plexus, in addition to noninvasive physical therapy. Ultimately the patients who elect to undergo brachial plexus decompression surgery are those in whom physical therapy or other pain management techniques fail, and they or a parent or guardian understand and consent to the risks of surgery.

Early Surgery May Result in Better Outcomes

Univariate analyses of factors that might be associated with an excellent surgical outcome, defined as pain relief of $\geq 90\%$, determined that patients who had excellent outcomes were significantly younger than those who did not. In addition, patients with excellent outcomes trended toward having shorter durations of preoperative symptoms. These 2 factors may be covariant, because patients who present for surgical assessment earlier in the course of their disease are likely to be younger. Taken together, these 2 factors suggest that expediting surgical evaluation may result in better long-term outcomes, perhaps because it decreases the duration of brachial plexus compression and forestalls the development of a chronic pain syndrome. Another possibility is that older children may have less neural plasticity and intrinsic regenerative abilities, in addition to a more severe or long-standing underlying neurological injury.

Pediatric Versus Adult nTOS: Different Mechanisms of Injury?

Neurogenic TOS is much less common in the pediatric population compared with adults; however, the incidence of cervical ribs in the pediatric nTOS population is much higher. In our cohort, 64.5% of nTOS cases were associated with a bony anomaly. This agrees with previous studies that demonstrate the rate of cervical ribs in the pediatric nTOS population to be approximately 53% (Table 5). In contrast, the incidence of cervical ribs in a recently published adult TOS series with 538 patients was 8.8%.¹⁵ This discrepancy suggests that the mechanisms of injury in adult nTOS and pediatric nTOS are divergent and may be more anatomical than functional in children. There may also be a mechanism by which nTOS can resolve spontaneously in adulthood after a period of growth or development in children with cervical ribs. Currently, we do not understand the natural history of pediatric nTOS sufficiently to predict how symptoms may change over long periods of time.

Adults with nTOS often report a history of trauma.⁶ In comparison, trauma was relatively rare in our pediatric cases (3 patients). However, nearly all of our patients played sports, many with an overhead motion component. A significant sports history was also noted in several other larger pediatric nTOS case series.^{4,26,28} This suggests that in adults, formation of scar tissue and direct injury to nervous structures may incite the development of nTOS, but in children, the underlying mechanical forces may relate

more to hypertrophy of muscles (particularly in a crowded region of the neck, and in the context of a cervical rib) and direct compression or entrapment of the plexus. Another hypothesis is that both kinds of injury result in a chronic inflammatory state that is uniquely detrimental to nerve tissue. Rehemutula et al. sent 4 lymph node biopsies for pathological analysis from the fat pad of the posterior cervical triangle, which appeared large; all 4 were found to have chronic inflammation.¹⁶ We do not routinely perform lymph node biopsies, but the 3 children who responded to US-guided steroid injections of the plexus in our series may offer evidence that antiinflammatory strategies are useful adjuncts to surgery. No studies have examined the utility of brachial plexus injections in nTOS, as either a diagnostic or therapeutic tool for children. Two studies describe anterior scalene muscle blocks to be predictive of successful surgical outcomes in nTOS in adults.^{1,10} We are currently studying the positive predictive value of these injections, but the lack of response is not a contraindication to surgery.

Limitations of the Study

The primary limitations of our study are its retrospective nature and nonblinded postoperative assessments. Patients are more likely to underreport negative outcomes when the questioner is their surgeon. To address these shortcomings, prospective, long-term observational trials with patient-reported quality of life measures need to be performed.

Conclusions

Pediatric nTOS is a rare pain syndrome that can be challenging to diagnose. Surgical decompression of the thoracic outlet results in complete pain relief in approximately one-half to two-thirds of patients, and $> 50\%$ pain relief in 90% of patients. Complications are rare but potentially serious, such as phrenic nerve injury and pneumothorax. Adult and pediatric nTOS may differ in pathogenesis and response to treatment.

References

1. Ambrad-Chalela E, Thomas GI, Johansen KH: Recurrent neurogenic thoracic outlet syndrome. *Am J Surg* **187**:505–510, 2004
2. Arthur LG, Teich S, Hogan M, Caniano DA, Smead W: Pediatric thoracic outlet syndrome: a disorder with serious vascular complications. *J Pediatr Surg* **43**:1089–1094, 2008
3. Çağlı K, Özçakar L, Beyazit M, Sirmali M: Thoracic outlet syndrome in an adolescent with bilateral bifid ribs. *Clin Anat* **19**:558–560, 2006
4. Chang K, Graf E, Davis K, Demos J, Roethle T, Freischlag JA: Spectrum of thoracic outlet syndrome presentation in adolescents. *Arch Surg* **146**:1383–1387, 2011
5. Dahlin LB, Backman C, Düppe H, Saito H, Chemnitz A, Abul-Kasim K, et al: Compression of the lower trunk of the brachial plexus by a cervical rib in two adolescent girls: case reports and surgical treatment. *J Brachial Plex Peripher Nerve Inj* **4**:14, 2009
6. Fugate MW, Rotellini-Coltvet L, Freischlag JA: Current management of thoracic outlet syndrome. *Curr Treat Options Cardiovasc Med* **11**:176–183, 2009
7. Hefford C, Abbott JH, Arnold R, Baxter GD: The patient-

- specific functional scale: validity, reliability, and responsiveness in patients with upper extremity musculoskeletal problems. **J Orthop Sports Phys Ther** 42:56–65, 2012
8. Hempel GK, Shutze WP, Anderson JF, Bukhari HI: 770 consecutive supraclavicular first rib resections for thoracic outlet syndrome. **Ann Vasc Surg** 10:456–463, 1996
 9. Huang JH, Zager EL: Thoracic outlet syndrome. **Neurosurgery** 55:897–903, 2004
 10. Jordan SE, Machleder HI: Diagnosis of thoracic outlet syndrome using electrophysiologically guided anterior scalene blocks. **Ann Vasc Surg** 12:260–264, 1998
 11. Khan A, Rattihalli RR, Hussain N, Sridhar A: Bilateral thoracic outlet syndrome: An uncommon presentation of a rare condition in children. **Ann Indian Acad Neurol** 15:323–325, 2012
 12. Martins RS, Siqueira MG: Cervical rib fracture: an unusual etiology of thoracic outlet syndrome in a child. **Pediatr Neurol** 43:293–296, 2007
 13. Maru S, Dosluoglu H, Dryjski M, Cherr G, Curl GR, Harris LM: Thoracic outlet syndrome in children and young adults. **Eur J Vasc Endovasc Surg** 38:560–564, 2009
 14. O'Brien PJ, Ramasunder S, Cox MW: Venous thoracic outlet syndrome secondary to first rib osteochondroma in a pediatric patient. **J Vasc Surg** 53:811–813, 2011
 15. Orlando MS, Likes KC, Mirza S, Cao Y, Cohen A, Lum YW, et al: A decade of excellent outcomes after surgical intervention in 538 patients with thoracic outlet syndrome. **J Am Coll Surg** 220:934–939, 2015
 16. Rehemutula A, Zhang L, Chen L, Chen D, Gu Y: Managing pediatric thoracic outlet syndrome. **Ital J Pediatr** 41:22, 2015
 17. Rochkind S, Shemesh M, Patish H, Graif M, Segev Y, Salame K, et al: Thoracic outlet syndrome: a multidisciplinary problem with a perspective for microsurgical management without rib resection. **Acta Neurochir Suppl** 100:145–147, 2007
 18. Roos D: Overview of thoracic outlet syndromes, in Machleder HI (ed): **Vascular Disorders of the Upper Extremity**. Mount Kisco, NY: Futura, 1989, pp 155–177
 19. Sanders RJ: Results of the surgical treatment for thoracic outlet syndrome. **Semin Thorac Cardiovasc Surg** 8:221–228, 1996
 20. Sanders RJ, Annest SJ, Goldson E: Neurogenic thoracic outlet and pectoralis minor syndromes in children. **Vasc Endovascular Surg** 47:335–341, 2013
 21. Sen S, Dişçigil B, Boga M, Ozkisacik E, Inci I: Thoracic outlet syndrome with right subclavian artery dilatation in a child – transaxillary resection of the pediatric cervical rib. **Thorac Cardiovasc Surg** 55:339–341, 2007
 22. Shepherd J: Thoracic outlet compression syndrome: a rare cause of upper extremity symptoms in children. **J Am Board Fam Pract** 12:403–405, 1999
 23. Stansby G, Lambert D: Thoracic outlet syndrome in children and young adults. **Eur J Vasc Endovasc Surg** 38:565–566, 2009
 24. Stratford PW, Binkley JM, Stratford DM: Development and initial validation of the upper extremity functional index. **Physiother Can** 53:259–267, 2001
 25. Swierczyńska A, Kłusek R, Krocza S: [Neurorehabilitation in children with thoracic outlet syndrome and its assessment.] **Przegl Lek** 62:1308–1313, 2005 (Polish)
 26. Teddy PJ, Johnson RD, Cai RR, Wallace D: Surgery for paediatric thoracic outlet syndrome. **J Clin Neurosci** 19:235–240, 2012
 27. Turgut M: Thoracic outlet syndrome in children. **J Pediatr Orthop** 17:697, 1997
 28. Vercellio G, Baraldini V, Gatti C, Coletti M, Cipolat L: Thoracic outlet syndrome in paediatrics: clinical presentation, surgical treatment, and outcome in a series of eight children. **J Pediatr Surg** 38:58–61, 2003
 29. Vu AT, Patel PA, Elhadi H, Schwentker AR, Yakuboff KP: Thoracic outlet syndrome in the pediatric population: case series. **J Hand Surg Am** 39:484–487, 487.e1–487.e2, 2014
 30. Yang J, Letts M: Thoracic outlet syndrome in children. **J Pediatr Orthop** 16:514–517, 1996

Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Hong, Ali, GG Heuer. Acquisition of data: Zager, Hong, Pisapia, Alexander, AJ Heuer. Analysis and interpretation of data: Hong, Ali, GG Heuer. Drafting the article: Hong, Pisapia. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Statistical analysis: Hong. Study supervision: Hong.

Correspondence

Eric L. Zager, Department of Neurosurgery, University of Pennsylvania, 3400 Spruce St., 3rd Fl. Silverstein Pavilion, Philadelphia, PA 19104. email: eric.zager@uphs.upenn.edu.