Thoracic Outlet Syndrome in Paediatrics: Clinical Presentation, Surgical Treatment, and Outcome in a Series of Eight Children

By G. Vercellio, V. Baraldini, C. Gatti, M. Coletti, and L. Cipolat
Milan, Italy

Background/Purpose: This report reviews the clinical presentation, surgical treatment, and outcome of 8 children treated for the thoracic outlet syndrome (TOS) during the last 3 years.

Methods: From 1998 through 2001 31 patients were admitted to our Vascular Surgery Unit with TOS. Eight of them (25.8%) were in the paediatric age group, 8 to 16 years (mean, 13 years). No sex prevalence was found. The presenting symptoms were neurologic in 2 patients (25%) and secondary to venous flow impairment in 6 (75%). At phlebography, venous thrombosis was seen in 2 cases, and functional intermittent obstruction was seen in 4. Seven patients underwent decompressive surgical partial resection of the first rib with transaxillary or supraclavicular access. One patient was treated conservatively.

Results: There were no major postoperative complications. Mean hospital stay was 2.7 days. In no patient were there signs of recurrence after a mean follow-up of 18 months (range, 3 to 36 months).

Conclusions: In the authors' experience TOS in paediatric patients occurs with the same symptoms and thrombotic complications as in adults. The same surgical strategy adopted in adult patients is advisable for affected children.

INDEX WORDS: Thoracic outlet syndrome, first rib resection.

THORACIC OUTLET SYNDROME (TOS) identifies the clinical condition determined by the mechanical compression and entrapment of the subclavian vessels and the brachial plexus cords within the space delimited by the scalene muscles, the clavicle, and the first rib. This compression can be secondary to anatomic bony anomalies (cervical rib, fibrous bands, mega-apophysis) or to hypertrophy of the anterior and middle scalene muscles, subclavius muscle, or pectoralis minor muscle.[1,2] Combined congenital anatomic anomalies and acquired functional factors (muscular hypertrophy or contracture) acting together determine the wide spectrum of vascular and neurologic symptoms that usually present during the third to fourth decade of life. From a recent review of the literature very few cases of TOS occurring in paediatric age and requiring surgical treatment have been reported. In the widest series, mean age of patients is about 35 years, and the youngest patients among the reported cases are 15 years old.[1,3-7]

This study reviews the clinical presentation, surgical treatment, and outcome in a series of 8 children treated for TOS during the last 3 years.

MATERIALS AND METHODS

Between December 1998 and December 2002, 31 patients were admitted to our vascular surgery unit with TOS. Eight of them (25.8%) were in the paediatric age range of 8 to 16 years (mean, 13 years). There were 4 boys and 4 girls.

At presentation, neurologic symptoms (paraesthesiae in the hand and pain in the neck) were present in 2 patients (25%) and correlated with compression by mega-apophysis or cervical rib on the brachial plexus documented at x-ray scan. Symptoms secondary to venous outflow impairment (cyanosis and oedema) were present in 6 patients (75%) who underwent investigation by dynamic plebography. Venous thrombosis was seen in 2 patients (Fig 1) and functional intermittent obstruction in 4 cases (Fig 2). Plebography was carried on with the involved arm in 90° abduction and external rotation. In 2 patients, TOS was associated with venous malformations of the same limb with congestive symptoms made worse by arm abduction. In all patients, preoperatively, the Wright manoeuvre and EAST (Elevated Arm Stress Test) described by Roos [8] were diagnostic, whereas the Adson test result was positive in only 2 cases. In only one (a 12-year-old girl basketball player) was the onset of symptoms related to traumatic events, occupational, or sports activities. No patient was investigated by arteriography or electromyography. The clinical findings are summarised in Table 1.

On the basis of the findings in 7 patients there was an indication for surgical decompression of the thoracic outlet. Four patients, suffering from venous symptoms, underwent scalenotomy and first rib partial resection with transaxillary access. Tenotomy of the pectoralis minor was performed in all cases. In 2 patients, with neurologic symptoms related to a cervical rib and mega-apohysis, scalenectomy, first rib partial resection, and excision of the bone anomalies were performed via a supraclavicular access. One patient, suffering from venous outflow impairment associated with major neurologic symptoms, under-

From the Vascular Surgery Unit, "V Buzzi," Children's Hospital, Milan, Italy.

Presented at the 49th Annual Congress of the British Association of Paediatric Surgeons, Cambridge, England, July 23-26, 2002.

Address reprint requests to Dr Vittoria Baraldini, Ospedale dei Bambini "V Buzzi," Via Castelvetro 32, 20154 Milano, Italy.

0022-3468/03/3801-0011$35.00/0
doi:10.1053/jpsu.2003.50010

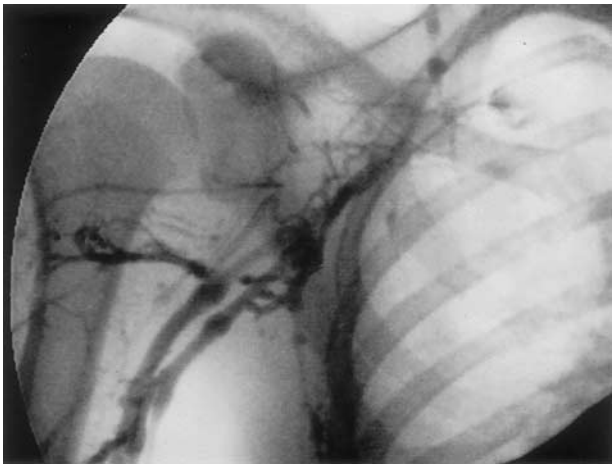

Fig 1. Dynamic phlebograpy: venous thrombosis of the axillary-subclavian vein with improved collateral circulation.

went scalenectomy, total first rib resection, and circumferential venolysis through a combined supra and infraclavicular approach.

One patient, a 14-year-old girl with the Klippel-Trenaunay syndrome in her left lower limb and both upper limbs, was treated conservatively because phlebography showed only a moderate impingement on the subclavian vein in the costoclavicular space.

RESULTS

There were no major postoperative complications in the 7 patients submitted to surgery. The postoperative hospital stay ranged from 1 to 7 days (mean, 2.7). In one patient a chest tube was placed for a pleural tear. One patient had transient numbness of the posterior aspect of the arm in the area of the intercostobrachial nerve. Two patients had transient paraesthesia in the area of the C8-T1 trunk.

All the patients presenting symptoms of venous outflow impairment (cases 2, 3, 4, 5, and 6) underwent a control dynamic phlebography 1 to 3 months after surgery to assess patency or residual entrapment of the vein. Minimal residual impingement was seen in one of the 4 patients with previous intermittent positional compression. One patient with vein thrombosis showed an early recanalization with rethrombosis and recurrence of symptoms 6 months later. A second patient with vein thrombosis showed at phlebography an improvement in collateral circulation.

All patients were submitted to a clinical reexamination. After a mean follow-up of 18 months (range, 6 to 36), 6 patients were symptom free, one patient conservatively treated still complained of minor discomfort, and one patient with recurrent thrombosis showed persistent nondebilitating oedema. The treatment strategies, complications, and outcome are summarised in Table 2.

DISCUSSION

The management of the Thoracic Outlet Syndrome is controversial. Diagnostic tools, conservative or surgical treatment, the best timing for surgery, surgical techniques, and the appropriate incisions are debated. Even the methods for assessing the effectiveness of treatment are controversial.[9] This syndrome is characterised by a constellation of neurologic and vascular symptoms, sometimes subjective, in a few cases evoked by traumatic events, work, or sporting activities.[6,7,10,11] Furthermore, psychologic and medicolegal aspects related to this syndrome can be misleading and interfere with the evaluation of results.[7,12] These can very well explain the reluctance of vascular and other specialist surgeons to take care of patients affected by this condition. A recent review of the literature found no publication dealing with incidence and management of this syndrome in children or adolescents.

This series of 8 children affected by TOS observed and treated over a 3-year period in our vascular surgery unit by one consultant surgeon shows that this syndrome is present in a significant number of children. Three more patients in the paediatric age group (not included in this series) were seen in the last 6 months further underlining the importance of this syndrome, which often is misdiagnosed in children.

The diagnosis of TOS is based on a careful evaluation of the symptoms and clinical examination. Frequently, patients complain of paraesthesia in the area of the C8-T1 roots (lower tract syndrome). Pain referred to the superclavicular area and headaches are associated commonly with the presence of anatomic bony anomalies such as mega-apophysis, cervical rib, or fibrous bands (upper tract syndrome).[8] In our series, 2 patients presented with neurologic symptoms in association with anatomic bony anomalies.

Oedema and cyanosis can be caused by intermittent impairment of venous flow or thrombosis of the axillary-subclavian venous tract secondary to compression of the vein within the costo-clavicular space or under the pectoralis minor muscle. The so-called subclavian vein "ef-

Fig 2. Dynamic phlebography: postural functional obstruction of the subclavian vein; involved arm in abduction and external rotation.

Table 1. Clinical Findings

Patient No.	Sex/Age (yr)	Delay to Onset	Side	Anatomic Anomalies	Prevailing Symptoms	Associated Symptoms
1	F/12	15 mo	Right	Mega-apophysis	Neurological	Arterial
2	M/8	15 d	Left	—	Venous	—
3	M/15	2 yr	Right	—	Venous	Neurological
4	F/14	1 mo	Right	—	Venous*	Arterial
5	M/11	1 yr	Left	—	Venous*	—
6	M/15	7 mo	Left	Venous malformation	Venous	—
7	F/14	1 yr	Left	Venous malformation	Venous	—
8	F/16	6 mo	Right	Cervical rib	Neurological	Arterial

*Thrombosis.

fort" thrombosis or Paget-Schroetter syndrome generates major disability with risk of pulmonary embolism.[13-15] In our series, 6 patients complained of symptoms secondary to impairment of venous flow; 2 of these had segmental venous thrombosis.

Many patients complain that simple everyday movements induce fatigue in the involved arm secondary to intermittent arterial compression. This symptom was present in 3 patients. Arterial lesions (thrombosis and aneurysms) are rare and almost always secondary to arterial trauma from a cervical rib.[16]

Regarding the proper physical examination, after a careful inspection and palpation of the supraclavicular fossae (for masses or abnormal pulsation), there is general agreement that among the classic provocative manoeuvres, the Adson test is less reliable than the Wright test (signs of arterial compression in hyperabduction) and the EAST (elevated arm stress test). The last, described by Roos, involves having the patient assume the extended arm position at 90° and rapidly flex and extend the fingers. The reproduction of symptoms is significant as a predictor of the success of the first rib resection.[8]

All the patients must have x-rays to detect bony anomalies (cervical rib, mega-apophisys, unusually large first rib or scalene tubercle, acquired or congenital clavicle pseudoarthrosis). Electrodiagnostic studies (electromyography and somatosensory evoked potentials) must be carried out in cases of suspected peripheral entrapment or in cases of suspected partial denervation of the intrinsic hand muscles.[17] No patients in our series required electrodiagnostic studies.

All patients with venous impairment must be submitted to dynamic phlebography because Colour-Doppler investigation can be unclear in the assessment of venous entrapment or thrombosis. It is agreed generally that conservative treatment by postural correction, stretching, and strengthening exercises is effective only in cases of early and minor symptoms.[13] Three of the patients operated on in our series had been treated previously by physiokinesitherapy for 3 months with no benefit.

In the case of venous thrombosis, the recommended treatment is thrombolysis followed by surgical decompression of the thoracic outlet.[14,18,19] In the 2 patients in our series affected by venous thrombosis, thrombolysis was not performed because at the time of diagnosis the thrombosis was of more than a week's duration. Both of them underwent surgery for first rib partial resection through the transaxillary approach. In cases of chronic/recurrent subclavian vein thrombosis, thoracic outlet decompression by first rib resection should improve the collateral venous circulation.[14]

To avoid pitfalls in diagnosis, extensive history taking and detailed clinical examination should be performed by experienced clinicians. The decompressive operation should be performed by a surgeon experienced in all surgical options.

Although there is no doubt about the effectiveness of first rib resection and the unreliability of simple scalenotomy, there is no general agreement about the best surgical approach for rib resection. The axillary route, popularised by Roos in 1965, seems to be the best technique to decompress the vein, allowing removal of the anterior segment of the rib. Several investigators perform this operation through the axillary approach as a

Table 2. Treatment Strategies

Patient No.	Approach	Complications	Postoperative Hospital Stay (d)	Follow-Up (mo)	Clinical Results
1	Supraclavicular	—	2	24	Asymptomatic
2	Transaxillary	—	2	6	Asymptomatic
3	Supra + infraclavicular	—	7	36	Asymptomatic
4	Transaxillary	Pnx	3	6	Asymptomatic
5	Transaxillary	—	3	6	Recurrence
6	Transaxillary	—	1	31	Asymptomatic
7	Physiokinesitherapy	—	—	12	Improved
8	Supraclavicular	—	1	22	Asymptomatic

first choice even in a case with associated cervical rib.[1,4,6,8] Other investigators prefer the supraclavicular approach because it is much easier, through this approach, to recognise and correct anatomic anomalies or arterial lesions; rib resection is limited to the posterior and middle segment.[7,13,14,20] It has been suggested recently that the first rib be resected totally through a double supra/infraclavicualar approach.[10,13,14] This approach permits a circumferential venolysis, and it is particularly indicated in case of established venous flow impairment.

The senior author, based on more than 200 operations performed from 1977, has developed a personal algorithm to choose which of the 3 incisional approaches should be used (axillary, supraclavicular and combined).[21] When anatomic bone anomalies (cervical rib or mega-apophysis) or arterial lesions (thrombosis or aneurysm) are present, it is essential to perform surgical decompression, the correction of anatomic anomalies, and deal with associated arterial lesions by a supraclavicular approach. This approach also is advisable in cases of neurologic signs of compression on C5-C6-C7 roots. In all the other cases (venous or arterial intermittent compression or neurologic symptoms secondary to compression on C8-T1 roots) the axillary incision is preferable for a better cosmetic result. To achieve better intraoperative exposure and a more complete decompression we add tenotomy of the pectoralis minor muscle. Finally, the combined approach is advisable in cases of complex symptomatology referable to recurrent impairment of venous flow in association with neurologic involvement of the upper brachial plexus cords.