Surgery Remains the Most Effective Treatment for Paget-Schroetter Syndrome: 50 Years' Experience

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Background. **Significant improvements were made in the diagnosis and management of Paget-Schroetter syndrome (thrombosis of the axillary–subclavian vein) secondary to thoracic outlet syndrome during the past 50 years. The diagnosis has often been extremely difficult.**

Methods. **Multiple approaches both in diagnosis and therapy have been tried during the years. After recognizing that the underlying pathologic process resulted from an abnormal insertion of the costoclavicular ligament laterally on the first rib, along with hypertrophy of the scalenus anticus muscle, 506 of 626 extremities have been managed by thrombolytic therapy followed by prompt transaxillary resection of the first rib. These patients have been followed up from 1 to 32 years (average of 7.2 years ± 1.0 standard deviation).**

Results. **Four hundred eighty-six patients (96%) improved. Because the pathophysiology is not well understood, many venograms suggest intraluminal disease rather than external compression. Therefore, attempts at opening the narrowed vein with intraarterial techniques do not work. Use of percutaneous venous angioplasty with stents have all occluded in our experience, making further management difficult. Venous bypass grafts fail because of low venous pressure.**

Conclusions. **Recognition that an abnormal congenital lateral insertion of the costoclavicular ligament on the first rib causes venous occlusion in Paget-Schroetter syndrome has led to acute thrombolysis, followed by prompt first rib resection, as the ideal management.**

(Ann Thorac Surg 2008;86:254–60)

Paget-Schroetter syndrome (PSS), or "effort" thrombosis of the axillary subclavian vein, is most often secondary to thoracic outlet syndrome (TOS). It usually occurs in patients with excessive arm activity in the presence of one or more compressive elements in the thoracic outlet. The syndrome was described independently by Von-Schroetter [1] in 1884 in Vienna and by Paget [2] in 1875 in London. For many years, therapy included elevation of the arm with anticoagulants and subsequent return to work [3, 4]. If symptoms recurred, the patient was considered for first rib resection with or without thrombectomy [5, 6]. When the congenital lateral insertion of the costoclavicular ligament was recognized as the underlying cause, prompt transaxillary rib resection was used immediately after thrombolysis.

The availability of thrombolytic agents [7–11], combined with prompt surgical neurovascular decompression of the thoracic outlet [5], has reduced morbidity and necessity for thrombectomy, substantially improving clinical results including the return to work. For those seen late (greater than 6 weeks after the thrombotic episodes), thrombolytic agents followed by first rib resection have been successful, but slightly less so.

The purpose of this report is to define the evolution of diagnosis and therapeutic management in patients with effort thrombosis secondary to TOS [12–15], based on understanding the anatomy and pathophysiology, and to compare it with the natural history of the disease [16–18].

The axillary subclavian vein traverses the tunnel formed by the clavicle and subclavius muscle anteriorly, the scalenus anticus muscle laterally, the first rib posterior-inferiorly, and the costoclavicular ligament medially (Fig 1).

In most patients with thrombosis of the axillary subclavian vein (PSS), the costoclavicular ligament congenitally inserts further laterally than normal (Fig 2). When the scalenus anticus muscle, which is lateral to the vein, becomes hypertrophied through activity and exercise, the vein is significantly narrowed. This is not the case when the costoclavicular ligament inserts in a normal place much more medially on the first rib, even with significant scalenus anticus muscle hypertrophy.

When the vein clots secondary to TOS and PSS evolves, there is usually a severe inflammation in the area of the thoracic outlet. This markedly handicaps the anatomic dissection by obscuring the location of the first rib and the anatomy of the thoracic outlet. In addition to the inflammation, which often plasters the neurovascular structures onto the chest wall obliterating the view of the

Accepted for publication March 7, 2008.

Presented at the Fifty-fourth Annual Meeting of the Southern Thoracic Surgical Association, Bonita Springs, FL, Nov 7–10, 2007.

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Published by Elsevier Inc

0003-4975/08/$34.00
doi:10.1016/j.athoracsur.2008.03.021

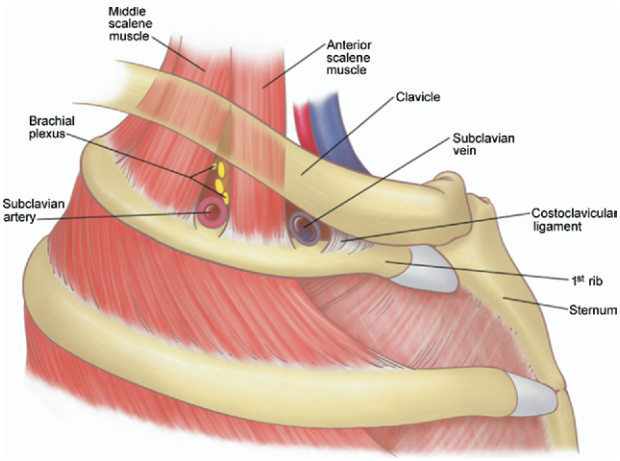

Fig 1. Normal anatomy of the thoracic outlet with conventional insertion of the costoclavicular ligament on the first rib.

first rib, there is a loss of the usual blue color of the vein because of the lack of blood flowing through it. This removes one of the best anatomic landmarks, further increasing the difficultly for the surgeon both in the acute and subacute phase of the syndrome.

Material and Methods

The chair of the institutional review board of the Baylor Research Institute reviewed the study and determined that it was exempt from institutional review board approval under 45 CFR 46.101 (b) (4).

Clinical manifestations of effort thrombosis of the axillary subclavian vein in the acute and subacute phases were evaluated in 626 extremities of 608 patients, 18 being bilateral. There were 307 women and 301 men, ranging in age from 16 to 51 years, with a mean of 32 years. (For the remainder of the discussion, the number

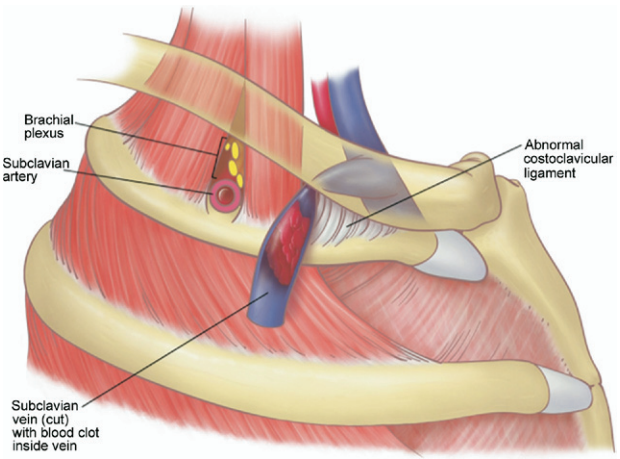

Fig 2. Congenital abnormal lateral insertion of the costoclavicular ligament on the first rib with hypertrophy of the scalenus anticus muscle lateral to the vein and thrombosis of the axillary-subclavian vein (Paget-Schroetter syndrome).

Table 1. Symptoms and Signs

Sign or Symptom	Number of Extremities
Venous distention (arm)	626
Subcutaneous venous collateral around shoulder (Urschel's sign)	618
Arm swelling	602
Bluish discoloration	591
Aching pain (with exercise)	207
Cervical ribs	62
Bilateral	18
Minimal symptoms	24

of the extremities will be assessed rather than the number of patients.) Four hundred thirty-two patients had unusual occupations that involved excessive, repetitive muscular activity of the shoulder, arm, and hand. Potentially aggravating occupations included such sports as golf, tennis, baseball, football, weight lifting, cheerleading, and drill team members, or other pursuits such as painters, beauticians, and linotype operators. The symptoms were usually exacerbated by working overhead, cold temperatures (weather or air-conditioning), or hanging the arm down for long periods.

Diagnosis

In 626 extremities, swelling or venous distention over the chest, arm, or hand occurred, suggesting the clinical diagnosis of venous obstruction (Table 1). Elevation of the arm or hand did not seem to change the configuration of the veins or swelling acutely. Bluish discoloration was observed in 544 arms, and aching pain, which was increased by exercise, occurred in 520. Cervical ribs were noted in 62 instances. The bilateral syndrome occurred in 18 patients (12 women and 6 men), simultaneously in 2 (1 with previous bilateral clavicular fractures), and sequentially in 16. In 24 patients, only minimal symptoms were present.

One hundred percent of the extremities demonstrated a positive Adson's sign, hyperabduction sign, or various other compressive signs related to the thoracic outlet compression. Diagnostic tests performed included venous ultrasound studies, venous scintillation scans, venography, plethysmography, temperature studies of the extremity, and bilateral upper extremities nerve conduction velocities including both the median and ulnar nerves.

The diagnosis was established by clinical history, physical examination, ultrasound studies, and venogram performed through a medial antecubital vein. An indwelling plastic catheter was inserted into the antecubital vein after 1980 so that serial venograms and thrombolytic injections could be performed.

Substantial narrowing or occlusion of the axillary subclavian vein in the area of the first rib and clavicle was observed in all patients on venogram. Some collateral circulation was evident in 621 extremities, although it was obviously not adequate if swelling was present. The