

## [ Orthopaedic Surgery ]

# Paget-Schroetter Syndrome: A Review of Effort Thrombosis of the Upper Extremity From a Sports Medicine Perspective

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Paget-Schroetter is a rare diagnosis in the general population; however, it is more common in younger, physically active individuals. Clinicians must be familiar with the symptoms, physical examination, and initial imaging and treatment to expedite care and prevent possible life-threatening complications. Urgent referral to a regional specialist may improve the opportunity for thrombolysis to restore blood flow through the subclavian vein and to decrease the chance of pulmonary embolus, recurrent thrombosis, or need for vein grafting, as well as to improve the time to return to full activity (athletics and/or manual labor).

**Keywords:** Paget-Schroetter syndrome; effort thrombosis; upper extremity; sports medicine

**T**horacic outlet syndrome (TOS) symptoms can be somewhat vague; the treatment is typically performed by vascular surgeons.<sup>39</sup> There are 3 distinct varieties; the most common is neurogenic, followed by venous and, finally, arterial. Paget-Schroetter syndrome (PSS), or effort thrombosis, is a subset of the venous subgroup. This syndrome was originally described in 1875 by Paget,<sup>29</sup> followed by von Schroetter's<sup>38</sup> theory in 1884. A history of venous obstruction with vigorous exercise or activity is present in approximately 60% to 80% of cases, with an incidence of 1 to 2 per 100,000 individuals per year.<sup>16,19,22,34</sup>

This condition occurs in baseball,<sup>10,26</sup> softball,<sup>28</sup> wrestling,<sup>24</sup> swimming,<sup>37</sup> hockey,<sup>7</sup> martial arts,<sup>41</sup> backpacking,<sup>20</sup> and billiards.<sup>17</sup> Four cases were confirmed over an 11-year period tracking 1 major league team and 1 collegiate baseball team.<sup>10</sup> A recent study identified 32 high-level athletes with effort thrombosis over a 10-year time frame, 14 of which were baseball players.<sup>26</sup> Overhead workers and manual laborers are considered "industrial athletes," subjecting their upper extremity to similar forces, which increases the likelihood of this condition. Clinicians should be aware of this condition and its symptoms because early intervention can be lifesaving.<sup>18</sup>

## ANATOMY/PATHOGENESIS

TOS can be caused by compression of the brachial artery and the brachial plexus passing through a triangular space created

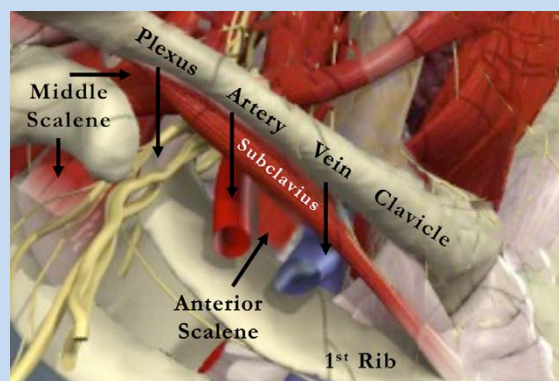


Figure 1. Clinical photograph of an 18-year-old man who presented to our clinic with 6 weeks of tightness in his right arm following throwing and weight lifting. He denied any pain or neurologic symptoms.

by the borders of the scalenus anticus muscle, scalenus medius muscle, and the first rib. The subclavian vein passes through a triangular space bordered by the first rib inferiorly, the scalenus anterior posteriorly, and the subclavius muscle and tendon medially just posterior to the intersection of the first rib and clavicle (Figure 1).

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Compression of the subclavian vein can occur with normal anatomy when the arm is in extremes of abduction and/or external rotation.<sup>2</sup> Compression occurs because of a hypertrophied scalenus anterior or subclavius, increased generalized muscle bulk, pectoralis minor or subscapularis muscles, coracocostal ligament, pervenous phrenic nerve, osseous exostoses or distorted bony anatomy (congenital or posttraumatic), or fibrous bands.<sup>31,39</sup> Interestingly, cervical ribs are more posterior structures, and while these may be the cause of neurogenic or arterial TOS, they are typically not the cause of venous TOS or effort thrombosis.<sup>33</sup>

Chronic compression of the vein leads to an inflammatory response in the soft tissues surrounding the vein, creating fibrotic connective tissue, which decreases the mobility of the vein.<sup>5</sup> As the arm moves, the vein is stretched, and small tears occur, causing further damage and predisposing it to thrombosis formation. Recurrent partial thrombosis followed by recanalization can also occur.<sup>2,23</sup> Over time, progressive intrinsic injury to the vein occurs, causing fibroelastic strictures, which creates turbulent blood flow and can lead to clot formation. Chronic compression may have relatively few, if any, symptoms until complete occlusion occurs, and even then, symptoms may be vague. Acute thrombosis typically results in more severe symptoms, such as severe swelling and pain, likely due to less collateral venous drainage.

## DIAGNOSIS

### Presentation

Patients with effort thrombosis usually present after the subclavian vein has completely thrombosed with a bluish hue to the arm, discoloration, asymmetric muscle bulk or increased postworkout swelling that does not dissipate, heaviness, and pain in the affected upper extremity (Figure 2). Symptoms typically occur within 24 hours of the heavy upper extremity activity, as superficial veins become prominent in the upper extremity as well as the neck. Effort thrombosis typically affects the dominant arm. In overhead athletes, complaints can mimic shoulder or elbow problems, causing loss of velocity or control, arm heaviness, or a “dead arm” (Table 1).

### Physical Examination

The asymmetric swelling is typically obvious through visual inspection. Superficial venous engorgement is present, and the axilla can be palpated for hard linear structures that represent the clotted vein. A careful neurologic and arterial examination should be performed on all patients with similar presenting symptoms to rule out neurogenic or arterial TOS.

Provocative testing for TOS pertains more to the neurologic or arterial subtypes. In the Adson test,<sup>4</sup> the head is turned toward the affected shoulder in slight extension and abduction while the patient inhales as the radial pulse is palpated.<sup>4</sup> Hyperabduction and external rotation while checking for paresthesia or changes in radial pulse are included in the Wright test<sup>40</sup> with both shoulders in 90° of abduction and



Figure 2. Venogram performed using conscious sedation and catheter placed in brachial vein. The venogram demonstrated an occlusion of the subclavian vein at the level of the clavicle/first rib with multiple collateral veins. A tPA infusion was performed; follow-up angiography noted resolution of thrombosis but residual stenosis due to scarring.

external rotation and the elbows flexed to 90°. In this position, the patient opens and closes the hands repeatedly, which may reproduce symptoms.<sup>30</sup> There is no specific physical examination finding that indicates effort thrombosis or PSS.<sup>32</sup> Imaging studies are the most reliable method of diagnosis.

### Imaging

The diagnostic study of choice is duplex ultrasonography. Sensitivity and specificity are excellent, ranging from 78% to 100% and 82% to 100%, respectively.<sup>9</sup> Positional venous occlusion may require the ultrasonographer to manipulate the arm to reproduce the vascular compromise. A fresh clot may not always be visualized because it may still be echolucent; a key diagnostic feature is the vein's lack of compressibility.<sup>19</sup> As the thrombus grows, it becomes more echogenic secondary to the fibrosis. In some cases, the large peripheral collateral veins may be present.

If ultrasonography is inconclusive, venography can be performed in the same setting as treatment (Figure 3). However, the diagnosis can be missed if the cephalic vein is used for contrast injection.

Table 1. Differential diagnosis of Paget-Schroetter

Neurologic thoracic outlet syndrome
Arterial thoracic outlet syndrome
Malignant tumors of the head, neck, or arm
Pancoast tumor
Complex regional pain syndrome
Brachial neuritis
Cervical nerve root compression
Quadrilateral space syndrome
Peripheral nerve compression

### Hypercoagulability Screening

The incidence of hypercoagulable conditions may be as high as 61% to 67%, especially in those without a clear anatomical cause of venous compression.<sup>8,10,13</sup> The initial treatment for thrombosis is the same regardless of the cause.

### CONSERVATIVE TREATMENT

Nonsurgical treatment using anticoagulation alone can result in acute pulmonary embolism in 6% to 15% and residual venous obstruction in over 75% of patients.<sup>3</sup> Outcomes are poor with the use of anticoagulation treatment as a single therapy with symptoms persisting in up to 91% of patients and up to 68% experiencing permanent disability.<sup>1,3,15,18,34</sup>

### OPERATIVE TREATMENT

Catheter thrombolysis with subsequent first rib resection has emerged as the treatment of choice for PSS.<sup>19</sup> Recanalization of the occluded vein is dependent on a number of factors.<sup>11</sup> The time from clot formation to treatment is a major factor. After 10 days, the success rate of opening the vein diminishes to almost zero.<sup>11,21,33</sup> Approximately 50% of veins treated at 6 weeks were partially opened, and none were completely opened.<sup>36</sup> A poor prognostic indicator is the size of the clot with longer thrombi correlating with poor success at recanalization.<sup>12</sup>

Thrombolysis is not a benign procedure; it can result in defects in the vein that predispose to repeat clot formation.<sup>27</sup> Angioplasty has not worked well because of the extrinsic nature of the compression, and venous stenting has resulted in stent fracture, deformation, and/or rethrombosis due to the unyielding surrounding structures.<sup>25</sup> Concurrent decompression of the anterior aspect of the first rib in the first few days following thrombolysis is recommended.<sup>19,36</sup>

Decompression can be performed by removal of the first rib and/or medial clavicle. Transaxillary resection produces



Figure 3. Diagram of the thoracic outlet, including the relationship of the subclavian artery and vein and brachial plexus to the bony and muscular architecture in the region.

excellent cosmesis with good to excellent results in 85% to 95% of patients.<sup>35</sup> Hemopneumothorax, long thoracic nerve injury, and incorrect rib removal can occur.<sup>19</sup>

### POSTOPERATIVE REHABILITATION/ RETURN TO PLAY

Anticoagulation for 3 to 6 months following recanalization and decompression is recommended for deep venous thrombosis.<sup>19</sup> Ultrasonography can monitor progress.<sup>19</sup>

In a recent study of 32 high-level athletes undergoing surgical management for venous TOS, 7 patients required repeat surgery (3 for bypass graft thrombosis, 2 for hemothorax, and 2 for wound hematoma or lymph leak).<sup>26</sup> The risk for recurrent thrombosis or reoperation was relatively high (22%), and the average return to play was 4.4 months. There was no difference in the return to play time based on level of athlete (high school vs collegiate vs professional) or sport.<sup>26</sup>

During anticoagulation, passive range of motion can be initiated to prevent shoulder stiffness. Strengthening was allowed by 1 month in several studies,<sup>10,14</sup> followed by a light toss program at 8 weeks, and interval throwing at 12 weeks.<sup>10</sup> The average time to return to sports was 3.5 months, with a mean return to full activity of 4.4 months.<sup>26</sup>

### CONCLUSION

PSS is rare. Immediate diagnosis can improve surgical outcomes and be lifesaving. The sooner treatment is initiated, the better the prognosis and the sooner the return to sport.

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