

Females With Subclavian Vein Thrombosis May Have an Increased Risk of Hypercoagulability

Kendall Likes; Danielle Rochlin, BA; Susanna M. Nazarian, MD; Michael B. Streiff, MD; Julie A. Freischlag, MD

Background: Subclavian vein thrombosis (SVT) is usually caused by vigorous activity or extensive use of the upper extremity. Patients are tested for hypercoagulability if they present with a spontaneous clot unassociated with such activity. The objective of this study was to determine the prevalence of hypercoagulability in patients undergoing first rib resection and scalenectomy presenting with SVT.

Methods: Using a prospectively maintained database from August 2003 through June 2011, patients were retrospectively reviewed for hypercoagulability testing and clinical outcomes.

Results: One hundred forty-three patients (79 females and 64 males; mean [range] age, 32 [16-71] years) presented with SVT, of whom 55 patients (43 females and 12 males; mean age, 32 [16-61] years) had undergone hypercoagulable testing. Fourteen patients (25.5%) (12 females and 2 males; mean age, 27 [16-46] years) had an abnormal hypercoagulable profile. A factor V Leiden

mutation was present in 6 patients, protein S deficiency in 4, a plasminogen-activator inhibitor-1 (PAI-1) deficiency in 2, and 1 patient each with protein C deficiency, anticardiolipin antibodies, factor VII mutation, factor II mutation, and antiphospholipid antibodies. Immediate and long-term postoperative vein patency was similar to patients without hypercoagulability. Patients were placed on lifelong anticoagulation therapy if they had a PAI-1, protein C, or protein S deficiency.

Conclusions: Patients with hypercoagulability do as well with first rib resection and scalenectomy for SVT as those without hypercoagulability. In our patient subset, more females were tested owing to a history of spontaneous thrombosis and an increased incidence of hypercoagulable disorders. Because of our findings, we believe younger women with SVT should undergo hypercoagulable testing to identify the need for long-term anticoagulation therapy.

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Author Affiliations: Division of Vascular and Endovascular Surgery and Medicine, Departments of Medicine (Dr Streiff) and Vascular and Endovascular Surgery (Mss Likes and Rochlin and Drs Nazarian and Freischlag) The Johns Hopkins Medical Institutions, Baltimore, Maryland.

PAGET-SCHROETTER SYNDROME was first used to describe stress thrombosis or spontaneous thrombosis of the subclavian or axillary vein in 1949 by the British surgeon Hughes.¹ Hughes coined the term after the British surgeon and pathologist, James Paget, and the Austrian internist and laryngologist, Leopold von Schrötter, who had previously described this phenomenon in the 1800s. Attributed to vigorous use of the upper extremity or a tight thoracic outlet, Paget-Schroetter syndrome is one of the consequences of thoracic outlet syndrome, or compression of the neurovascular bundle as it exits the chest. Anatomical causes of thoracic outlet syndrome include a wide lateral insertion of the costoclavicular ligament, hypertrophy or lateral insertion of the anterior scalene muscle, a cervical rib, or tightening of the space between the clavicle and first rib.

However, these anatomical abnormalities or activity risk factors are not identified in a subset of patients with Paget-Schroetter syndrome. Because venous thromboembolism is thought to develop as a consequence of abnormalities in blood flow, the vessel wall, and/or the coagulation potential of blood, one or more of these contributing factors presumably precipitates upper-extremity thrombosis in this subset of patients.

The objective of this study was to examine the demographic and clinical characteristics of this patient subset with Paget-Schroetter syndrome and determine the prevalence of hypercoagulability in patients undergoing first rib resection and scalenectomy (FRRS) who presented with subclavian vein thrombosis (SVT).

METHODS

A retrospective review of a prospectively maintained database from The Johns Hopkins Medi-

cal Institutions, Baltimore, Maryland, was performed. This database includes demographic information, initial presentation, patient history, prior treatments, hypercoagulable workup, surgical complications, and follow-up information regarding all patients who have undergone FRRS for thoracic outlet syndrome (TOS). The database has approval from the institutional review board of The Johns Hopkins Medical Institutions to follow the outcomes of these patients. Any additional information was obtained from the electronic patient records.

A total of 423 patients had undergone surgical intervention for TOS between August 4, 2003, and June 13, 2011, at either The Johns Hopkins Hospital or The Johns Hopkins Bayview Medical Center. A subset consisting of 143 of these patients was classified as presenting with Paget-Schroetter syndrome. Patients were included in the study if they presented with vascular compromise due to Paget-Schroetter syndrome, underwent FRRS, and returned for a venogram 2 weeks postoperatively. Patients who underwent FRRS but failed to return postoperatively and at 6-month intervals for routine duplex scans were excluded from this study. Patients who underwent surgical intervention for TOS prior to the inclusion of routine venography postoperatively were also excluded. No patients had central venous catheters placed in the past or had a diagnosis of malignant neoplasms.

Some patients presenting with Paget-Schroetter syndrome do not fall inside the traditional phenotype and medical history for the condition. Specifically, patients who are referred to us may lack a history of repetitive upper-extremity use, athletics, heavy lifting, or evidence of upper-body muscle hypertrophy. Hypercoagulability testing was performed preoperatively or postoperatively in these patients. The following thrombophilic conditions were assessed in 55 patients using standard laboratory techniques: factor V Leiden genotype, activated protein C resistance, prothrombin gene mutation genotype, protein C activity/antigen level, protein S activity/antigen level, antiphospholipid antibody syndrome, and plasminogen-activator inhibitor-1 (PAI-1; OMIM 613329) genotype. Patients were not screened for rarer hypercoagulable disorders, such as antithrombin III deficiency, or for thrombophilic conditions, such as homocysteinemia, that were not considered important factors for thrombosis in this patient subset.

Patients were evaluated preoperatively at The Johns Hopkins Medical Institutions for venous TOS. All the patients received venous TOS care according to standard Johns Hopkins Hospital protocol, regardless of preoperative intervention—transaxillary FRRS followed by venogram 2 weeks postoperatively. Postoperative anticoagulation therapy was prescribed to patients with confirmed hypercoagulable disorders and/or effort thrombosis. Three days following FRRS, all the patients were given subcutaneous low-molecular-weight heparin bridged to warfarin sodium. All the patients underwent venography 2 weeks after surgery to assess subclavian vein patency. The duration of anticoagulation therapy was determined after consultation with the hematology service as well as consideration of subclavian vein patency. If the subclavian vein was patent, no dilatation was done and anticoagulation therapy was stopped, if the patient did not have a coagulation abnormality. Subclavian veins that were more than 50% stenotic, as detected by venography, were dilated with balloon angioplasty and patients were given anticoagulation therapy until the vein showed good velocities in abduction and adduction using a duplex scan. Patients with chronically occluded subclavian veins, as seen on venography, were given anticoagulation therapy for up to 6 months after their surgery or until subsequent duplex scans revealed a recanalized subclavian vein. Patients with recurrent thrombosis after the postoperative venogram were also given anticoagulation therapy for up to 6 months or until the vein recanalized. Follow-up testing

Table 1. Characteristics of Patients With Hypercoagulable Syndromes

Patient No./Sex/Age, y	Occupation/Potential Predisposing Activity	Hypercoagulable State
1/M/46	Business/none indicated	Factor II mutation
2/F/29	Swimming while dehydrated	Factor V Leiden; PAI-1 deficiency
3/F/16	Student/none indicated	Factor V Leiden
4/F/34	None indicated	Factor V Leiden
5/F/19	Student/blow drying hair	Protein S deficiency
6/F/33	None indicated	Anticardiolipin syndrome
7/F/25	Student-athlete/weight lifting	Protein S deficiency
8/F/28	Heavy backpack while hiking	PAI-1 deficiency
9/F/30	None indicated	Factor V Leiden
10/F/17	Student/none indicated	Factor V Leiden
11/F/36	None indicated	Protein S deficiency; factor VII mutation
12/F/18	Student-athlete/dehydrated from running	Factor V Leiden
13/M/16	Student/wood chopping	Protein S deficiency
14/F/27	None indicated	Protein C deficiency/antiphospholipid syndrome

Abbreviation: PAI-1, plasminogen-activator inhibitor-1.

was performed at all postoperative visits and included subsequent duplex scans to assess vein patency. Successive duplex scanning allowed the identification of changes in veins over time. Patients who demonstrated good flow through the subclavian vein and adequate flow velocities in abduction, indicating minimal venous compression, stopped anticoagulation therapy if they did not carry a hypercoagulable diagnosis warranting lifetime anticoagulation therapy. Lifelong anticoagulation therapy was started if the patients were found to have protein C, protein S, or PAI-1 deficiencies or antiphospholipid syndrome. For those patients carrying a factor V Leiden deficiency, anticoagulation was stopped after 3 months, unless the patient also presented with a pulmonary embolism, in which case anticoagulation therapy was continued for 6 months.

RESULTS

Of the patients diagnosed with venous thoracic syndrome between August 4, 2003, and June 13, 2011, at The Johns Hopkins Medical Institutions, 143 patients presented with SVT. Of these patients, 55 had undergone hypercoagulable testing (43 females and 12 males; mean age, 32 [age range, 16-61] years).

Of those tested, 14 patients (25.5%) (12 females and 2 males; mean age, 27.7 [age range, 16-46] years) had an abnormal hypercoagulable profile. The characteristics of these patients are described in **Table 1**. Patients were diagnosed by history of activity when the throm-

Table 2. Comparison of Characteristics of Hypercoagulable vs Nonhypercoagulable Patients With Paget-Schroetter Syndrome

Characteristic	Hypercoagulable Syndrome (n = 14)	No Known Hypercoagulable Syndrome (n = 41)	P Value
Women, No. (%)	12 (85.7)	31 (75.6)	.40
Age, mean (SE), y	26.7 (2.38)	33.6 (2.01)	.04
Age of women, mean (SE), y	26.0 (2.02)	26.7 (2.38)	.007

bosis occurred. Any activity that may have prompted the thrombosis or any activity that had been repeated daily for years leading to the thrombosis was identified as a potential cause for upper-extremity deep venous thrombosis (DVT). Six patients were heterozygous for factor V Leiden, 4 had a protein S deficiency, 2 had a PAI-1 deficiency, 1 had antiphospholipid syndrome, 1 had anticardiolipin syndrome, 1 had a protein C deficiency, 1 had a factor VII mutation, and 1 had a factor II mutation. Seven of these patients underwent intervention, such as thrombolysis and/or angioplasty, prior to their FRRS operation to resolve venous thrombosis.

Comparison of patients with and without known hypercoagulability syndromes reveals a higher proportion of women in the hypercoagulable group (Table 2). Of the 12 males tested, 2 (17%) had an abnormal thrombophilia workup. Of the 43 women tested, 12 (29%) had an abnormal hypercoagulable workup. Of those who tested positive for carrying a thrombophilic syndrome, 2 (14%) were males and 12 (86%) were females. Patients presenting with venous thrombosis with thrombophilic conditions were also significantly younger than patients with Paget-Schroetter syndrome without known hypercoagulable disorders (mean [SE], 26.7 [2.38] vs 33.6 [2.01] years, $P = .04$). Of those tested for hypercoagulability, 7 patients presented with pulmonary embolism found incidentally, in addition to upper-extremity DVT, of which 1 tested positive for carrying a thrombophilic syndrome. Ten patients (71.4%) with a known hypercoagulable syndrome showed venous compression by duplex scan on diagnosis, compared with 27 patients (66%) in the subset without a known hypercoagulable disorder. Two patients (16.7%) with a hypercoagulable syndrome and 4 patients (12.9%) not carrying a known thrombophilic disorder were taking oral contraceptives at the onset of venous thrombosis.

At the routine 2-week postoperative venogram, 9 of the hypercoagulable patients were found to have stenotic veins, 1 patient had an occluded subclavian vein, and 4 patients had widely patent veins. The patients with stenotic veins underwent dilatation and were anticoagulated until the vein showed good velocities in abduction and adduction using a duplex scan. The average time of anticoagulation for patients that required postoperative dilatation was 2 months (range, 1-4 months). The 1 patient with an occluded vein was treated with anticoagulation therapy until the vein recanalized months postoperatively. There was no dif-

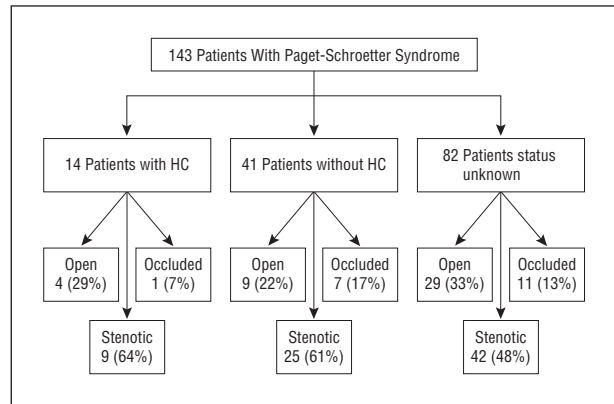


Figure 1. Subclavian vein status at 2-week postoperative venogram. HC indicates hypercoagulability syndrome. The remaining 6 patients did not have a venogram at 2 weeks after their surgery or the status of the venogram was unknown.

ference in outcomes detected at the 2-week postoperative venogram between the hypercoagulable and nonhypercoagulable groups (Figure 1). Two patients who tested for thrombophilia had recurrent thrombosis after their postoperative venogram and received anticoagulation therapy for 6 months. Both patients' veins recanalized, as seen using a duplex scan. In patients not tested for hypercoagulability who underwent surgical intervention for venous TOS, recurrent thrombosis also occurred in less than 5% of patients.

Patients in the hypercoagulable group were followed up with duplex scans for an average of 25.2 months (range, 8-73 months). Long-term patency, as seen using a duplex scan, was achieved in all 14 patients (100%). Patients with no known hypercoagulable syndrome were followed up an average of 15.6 months (range, 3-42 months) using a duplex scan; similar results were achieved using our protocol. At the time of their last duplex scan, all but 3 patients (93%) had open subclavian veins. All the patients continue to be evaluated regularly in the clinic as needed. Venography is performed at 2 weeks postoperatively so that therapeutic venoplasty can be performed if needed. But, because vein patency can be followed up using a duplex scan that has yielded excellent results, the use of multiple venograms is unwarranted in this patient subset because of the cost, risk, and pain associated with them.

COMMENT

In the 143 patients presenting with Paget-Schroetter syndrome and treated at The Johns Hopkins Medical Institutions, 14 patients (9.8%) were found to have at least one hypercoagulable disorder. The incidence of hypercoagulability in those not operated on remains unknown. Proper treatment of patients with a thrombophilic condition requires close coordination with a hematology service to ensure correct diagnosis and anticoagulation therapy. Awareness and proper management of their thrombophilic condition may prevent these patients from experiencing other thrombotic events, such as Paget-Schroetter syndrome recurrence, DVT, pulmonary embolism, or miscarriages. First-time venous throm-

boembolism is estimated to occur in 100 per 100 000 persons annually in the United States and has been shown to increase exponentially with age. At an average age of 32 years, one would expect fewer than 30 cases of venous thromboembolism per 100 000 persons.^{2,3} Hence, recognition of hypercoagulable syndromes in patients presenting with idiopathic Paget-Schroetter syndrome may prevent subsequent morbidity and/or mortality from venous thromboembolism.

In our series, patients diagnosed with a hypercoagulable disorder were more likely to have an idiopathic presentation, with no history of rigorous upper-extremity activity. A few patients in our study fell inside the typical active group and had a hypercoagulable disorder. There was a higher proportion of women than men with a hypercoagulable disorder. The hypercoagulable group was also significantly younger with a mean age of 27 (age range, 16-46) years compared with 32 (age range, 16-61) years for the nonhypercoagulable group ($P < .05$). Outcomes in postoperative venogram and in long-term vein patency did not differ between those with and without diagnosed hypercoagulable disorders.

Ours is not the first study to call attention to the coincidence of Paget-Schroetter syndrome and hypercoagulable disorders. Cassada et al⁴ retrospectively reviewed the outcomes of 18 patients they had treated for venous thoracic outlet syndrome between 2002 and 2004. There were 11 females and 7 males between the ages of 19 and 60 in this series. Twelve patients presented with acute venous thrombosis and 6 had swelling with thrombosis. All 18 patients underwent FRRS using a paracervical approach. All 12 patients who presented with acute thrombosis underwent thrombophilia testing and 8 (67%) were found to have at least one positive result. The following abnormalities were found: PAI-1 deficiency (5 patients), homocysteine (2 patients), anticardiolipin (2 patients), lupus anticoagulant (1 patient), and protein S deficiency (1 patient). Unlike our review, more complications occurred in those patients with thrombophilia that included hemothorax, recurrent thrombosis, and recurrent edema. Based on these findings, the authors recommended hypercoagulable testing in all the patients being considered for surgical intervention due to the risk of future thrombotic events. As is the case in this study, most previous studies have included even fewer patients than our series of 14.⁵⁻⁹

Héron et al¹⁰ evaluated 51 consecutive patients who presented with upper-extremity DVT without a history of central venous catheterization. There were 21 males and 30 females with a mean age of 32 (age range, 15-86) years. Twenty patients had a history of strenuous activity whereas 31 did not and were classified as having idiopathic disease. Those patients with hypercoagulable disorders were older (35 vs 28 years), were more likely to be female (81% vs 23%), and had a family history of thromboembolism (42% vs 15%). In addition, those idiopathic patients also had a higher chance of having a coagulation abnormality (42% vs 15%). Therefore, the authors concluded, similar to our findings, that those with idiopathic upper-extremity DVT especially with a family history of thromboembolism

should undergo hypercoagulable testing. However, unlike this study, our referral pattern is those primarily with effort thrombosis and not with hematological problems or cancer, which can account for the differing age distribution.

The literature seems to differ on the prevalence of hypercoagulable disorders associated with primary upper-extremity DVTs. Our conclusions are similar to those of a frequency-matched case-control study of patients with primary DVTs of the upper extremity. Martinelli et al¹¹ reviewed 36 patients who presented with primary DVTs of the upper extremities and compared their hypercoagulable profile to 121 patients who had primary DVTs of the lower extremities and to 108 healthy controls. Prevalence of abnormalities of the anticoagulation system and hyperhomocysteinemia was similar in those patients with an upper-extremity DVT and the control group (9% vs 6% and 6% vs 7%), respectively. Those with lower-extremity DVTs had a higher prevalence of 31% and 14%, respectively. Strenuous exercise was identified as the triggering event for thrombosis in 33% of those with upper-extremity DVT, whereas oral contraceptives, pregnancy, surgery, and immobilization were triggers for lower-extremity DVTs. Therefore, due to the low prevalence of hypercoagulable states found in patients with primary upper-extremity DVTs, the authors did not recommend routine screening for those presenting with upper-extremity DVTs.

Ruggeri et al¹² also found a low prevalence of thrombophilic coagulation defects in patients with upper-extremity DVTs. They described 27 consecutive patients (16 men; 11 women) with a mean age of 38 years (age range, 17-71 years). No central catheters had been placed in any of these patients. Two-thirds of the patients (18 of 27) had a history of strenuous activity prior to the subclavian or axillary vein thrombosis. Only 2 patients (7.4%) had abnormalities found in the hypercoagulable testing—one patient with protein C deficiency and the other with a factor V Leiden mutation. Four patients (14.8%) were found to have elevated anticardiolipin antibodies. No patient, however, underwent FRRS in this series.

In contrast, a more recent study reported a high incidence of coagulation abnormalities in patients with primary upper-extremity DVT. Hendler et al¹³ reviewed 31 patients (17 women; mean age, 38.8 years [age range, 16-60 years] and 14 men; mean age, 31.4 years [age range, 20-56 years]) who presented with upper-extremity DVT between 1993 and 2002. Fifteen were thought to have effort as their cause and 16 were described as spontaneous without a precipitating event or activity. There was no significant difference in the incidence of hypercoagulable factors. In the effort group, 53% of the patients were found to have at least one thrombophilic defect, as did 68.7% of the spontaneous group. These included the lupus anticoagulant (10 patients), anticardiolipin antibodies (4 patients), factor V Leiden (4 patients), prothrombin G20210A gene variant (3 patients), methylenetetrahydrofolate reductase (3 patients), and protein S deficiency (1 patient). Consequently, the authors recommend that all the patients presenting with primary upper-extremity DVT undergo hypercoagulability testing.

One concern in the promotion of screening for these disorders lies with the high costs of hypercoagulability testing. The complete battery of tests used in our study costs around \$850 (**Table 3**). Given this expense, we advise conducting these tests primarily on those patients whose Paget-Schroetter syndrome is believed to be without precipitating physical cause with low levels of upper-extremity activity, women, patients of small stature, and those without predisposing risk factors (pregnancy, dehydration, extensive travel, and others). We believe that the correlation between young small females is significant, as the thoracic inlet is smaller and more prone to thrombosis if a hypercoagulable state or disorder is present. With these findings, we can better predict groups of patients at risk for hypercoagulability. Because of the low incidence in certain patient subsets, expensive testing need not be performed routinely.

Table 3. Costs of Routine Hypercoagulable Tests^a

Test	Cost (US \$)
Antiphospholipid syndrome	225.28
Antithrombin activity	35.67
Factor V Leiden	135.18
PAI-1	93.87
Protein C antigen	112.64
Protein C activity	112.64
Protein C resistance	112.64
Protein S activity	112.64
Prothrombin gene mutation	135.18
Total	850.46

Abbreviation: PAI-1, plasminogen-activator inhibitor-1.
^aThe Johns Hopkins Medical Laboratories, 2012.

Chang et al¹⁴ showed the importance of routine venography postoperatively to direct treatment for patients undergoing FRRS for chronic subclavian vein thrombosis. Eighty-four patients were followed up using duplex scans for a mean (range) of 17.5 (2-69) months and long-term vein patency was evaluated. As seen by postoperative venogram, 21 patients had widely patent veins, 47 patients had stenotic veins requiring dilatation, and 16 patients had occluded veins requiring long-term anticoagulation therapy. In this series, 2 patients had late occlusions and symptomatic restenosis was seen in 3 patients. Long-term patency, as seen using a duplex scan, was achieved in 79 patients (94%) using this protocol. In our series, similar results were achieved—100% patency for the hypercoagulable group and 93% patency for those patients with no verified hypercoagulable disorder.

We propose the following algorithm for treatment of patients presenting with Paget-Schroetter syndrome and an atypical medical history (**Figure 2**). The algorithm rests on a high degree of suspicion for those with atypical presentation, coupled with close attention to results of the 2-week postoperative venogram, and a multidisciplinary approach with the hematology service and physical therapy.

In conclusion, hypercoagulability is a rare finding in those patients who present with Paget-Schroetter syndrome due to effort. The possibility of a hypercoagulability disorder should be considered in younger patients who present with lack of activity leading to the thromboses, especially if they are female. Consideration of this condition and judicious testing may yield improved outcomes and prevent future venous thromboembolic events in this subset of patients.

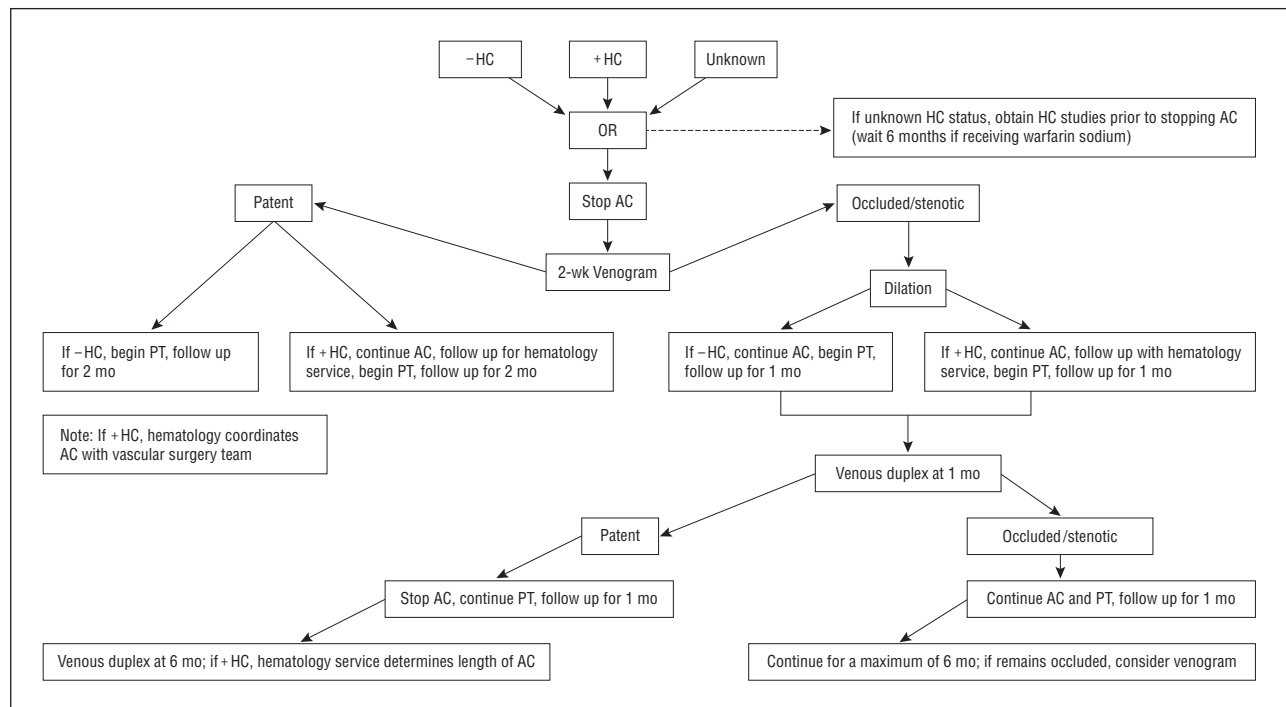


Figure 2. Treatment algorithm for patients with “idiopathic” Paget-Schroetter syndrome. AC indicates anticoagulation; HC, hypercoagulability syndrome; OR, thoracic outlet syndrome operation; PT, physical therapy; +, present; -, absent.

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Correspondence: Julie A. Freischlag, MD, Department of Vascular and Endovascular Surgery, The Johns Hopkins University, Ross Research Bldg, Ste 759, 720 Rutland Ave, Baltimore, MD 21205 (Jfreisc1@jhmi.edu).

Author Contributions: *Study concept and design:* Likes, Rochlin, Nazarian, Streiff, and Freischlag. *Analysis and interpretation:* Likes, Rochlin, Nazarian, and Freischlag. *Acquisition of data:* Likes, Rochlin, Nazarian, and Freischlag. *Drafting of the manuscript:* Likes, Rochlin, Nazarian, and Freischlag. *Critical revision of the manuscript for important intellectual content:* Likes, Rochlin, Nazarian, and Freischlag. *Study supervisor:* Freischlag.

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Additional Information: Ms Likes is an undergraduate student at Haverford College, Haverford, Pennsylvania. Ms Rochlin is a medical student at The Johns Hopkins Medical Institutions, Baltimore, Maryland.

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