INTRODUCTION
Thrombosis of the central veins is a rare disorder both in the lower and upper extremities. The annual incidence of upper extremity deep vein thrombosis (UEDVT) is 1/100,000 to 2/100,000 and accounts for only 1% to 4% of all patients presenting with DVT. The incidence of inferior vena cava (IVC) thrombosis as an isolated entity is unclear, with few reports in the literature. However, it is clear that IVC thrombosis is associated with a higher incidence of post-thrombotic syndrome (PTS). Many UEDVT and IVC thrombosis cases are secondary, related to the placement of long lines and catheters, pacemaker leads, and filters. This frequently adds to the complexity of treatment since additional considerations are required in planning the most appropriate intervention.

UPPER EXTREMITY CENTRAL VEIN THROMBOSIS
Primary UEDVT is mostly associated with the presence of Paget-Schroetter syndrome, which is defined as a DVT occurring secondary to anatomical abnormalities; this accounts for approximately 20% of all UEDVT patients. It may be known by various terms, such as “effort-related thrombosis” due to its close association with exercise as the stimulating event, and is principally related to compression of the thoracic outlet. The vast majority of cases, however, are secondary, resulting from intravenous catheter or pacemaker lead placement as well as malignant compression.

Involvement of the superior vena cava (SVC) in primary thrombosis is rarely reported and usually represents extension of thrombus from the upper limb. Although SVC thrombosis was originally thought to be related to malignancy, reports now suggest that benign causes may account for up to 40% of cases. This proportion is likely related to the increased frequency of central catheter access and pacemaker placement in the last several years.

It is clear that whatever the mechanism of UEDVT, PTS is a frequent complicating factor, and much work is still required to improve the diagnosis, classification, and standard of treatment.

ENDOVENOUS TREATMENT OF SVC THROMBOSIS AND UEDVT

SVC Thrombosis

There has been a sustained shift toward endovenous techniques, such as stenting and balloon angioplasty, for the treatment of SVC obstruction in the last several years. Although open surgical techniques have demonstrated good patency results over the long term, they have been performed in only a few specialist centers and have shown significant morbidity. Endovenous techniques have demonstrated similar patency rates, although there is a paucity of long-term follow-up data when compared to open surgery.

Treating chronic occlusion of the SVC carries significantly more potential for harm compared with a similar lesion in the IVC, where the risk of cardiac tamponade or hemothorax is not present. Most authors would therefore advocate performing these procedures in collaboration with cardiothoracic surgery in case it becomes necessary to open the chest. The risk increases with the complexity of the lesion, and type IV SVC obstructions (Table 1) are significantly less likely to be successfully treated.

There is no documented standard approach to endovenous treatment of SVC thrombosis, and a number of different techniques have been applied. In all cases, the operator needs to pay particular attention to the wire crossing to ensure that the wires do not enter the pericardium. Frequent views from both anterior-posterior, oblique, and lateral projections should be obtained to ensure that the wires cross in an appropriate plane. Similarly, post-crossing balloon dilation should be approached with caution, and the SVC should be dilated.
sequentially to minimize the potential for rupture. Post-dilation stenting of the SVC (as with all venous lesions) is likely to provide better long-term results and prevent early recoil of lesions. Good results have been obtained with a policy of stenting, although long-term outcome data has not yet been published (Figure 1).

**Type I:** Partial obstruction (up to 90% stenosis) of the SVC with patency of the azygos vein

**Type II:** Near complete obstruction (90-99%) of the SVC with patency and antegrade flow in the azygos vein

**Type III:** Near complete obstruction (90-99%) of the SVC with retrograde flow in the azygos vein

**Type IV:** Occlusion of the SVC with the involvement of one or more of the major caval tributaries and azygos vein

**Table 1.**
Stanford and Doty venographic classification of superior vena cava (SVC) obstruction.

---

**Paget-Schroetter Syndrome/Effort-Related Thrombosis**

Paget-Schroetter syndrome is associated with repetitive, strenuous activity of the upper extremities that ultimately compresses the thoracic outlet. The anatomical abnormalities at the costoclavicular junction that restrict the thoracic outlet are formed by the first rib, clavicle, and scalenus anterior muscle; there is also contribution from other abnormalities, such as the presence of a cervical rib, congenital bands, hypertrophy of scalenus tendons, and abnormal insertion of the costoclavicular ligament. The role of other factors such as hematological abnormalities, while undoubtedly part of the process, is less established.

Anticoagulation alone is still viewed by many as the gold standard for treating both upper- and lower-limb DVT. Clinical guidelines from the National Institute for Health and Care Excellence (NICE) in the United Kingdom recommend the use of direct oral anticoagulants, rivaroxaban, dabigatran, apixaban, or edoxaban as options for treating venous thromboembolism and preventing long-term recurrence. However, advances in catheter-directed lysis techniques and the introduction of pharmacomechanical techniques have highlighted that the long-term consequences of conservative management in a young patient population are unacceptable.

---

**Figure 1.**
A 63-year-old man with superior vena cava reconstruction with the WALLSTENT (Boston Scientific Corp.) following obstruction secondary to a hemodialysis line. (A) Pre-stent venogram, (B) pre-stent dilatation with balloon, and (C) post-stent venogram.
The accepted premise that intervention confers better results is based on the principle of restoring normal venous outflow from the upper limb. This ultimately involves two options for treatment: (1) venous lysis without surgery (e.g., lysis, venoplasty with or without stent placement), and (2) venous lysis plus surgery (with or without venoplasty and/or stent placement). Published outcomes for the first strategy report good results in up to 75% of patients, with the remainder requiring surgery at a delayed interval because of residual symptoms, mainly recurrent thrombosis and development of PTS. Venoplasty and stenting without rib resection was associated with stent fracture. Outcomes reported for the first strategy, thrombolysis followed by immediate surgery, demonstrated 100% long-term vein patency (6 months to 25 years follow-up), and all patients resumed normal activities following surgery. A strategy of selective surgical intervention after lysis, although sensible in principle to avoid the risks of surgery, is only possible with a robust care pathway that identifies which patients require surgery. However, no such pathway currently exists.

Thus, according to current evidence, the best treatment results for Paget-Schroetter syndrome are achieved by a policy of catheter-directed lysis followed by immediate correction of the anatomical abnormality (first rib resection) in all patients who are suitable for surgical intervention.

Line- and Catheter-Related Thrombosis

The risk of long-term complications in secondary UEDVT is lower than that observed in primary UEDVT, but significant complications may still occur. Therefore, the first-line strategy continues to be anticoagulation, with endovenous options limited to those patients who have a deteriorating course in the acute phase or develop symptomatic chronic disease.

In acute patients with severe symptoms, low bleeding risk, and long life expectancy, treatment should include catheter-directed thrombolysis or pharmacomechanical thrombectomy similar to the strategy for patients presenting with primary UEDVT.

In patients with a functioning catheter and stable symptoms, the catheter may be left in place while the patient is started on anticoagulation. Removing the lines and gaining access in alternative sites should only be considered for patients whose symptoms deteriorate.

Stenting in the upper extremities remains controversial; however, there are increasing reports of stent placement in patients whose cardiac pacemaker wires have caused significant obstruction (Figure 2). In these patients, it is better to remove the pacemaker wires, reconstruct the vessel (with or without removal of the first rib), and then replace the pacing wires. It is not recommended to simply stent the pacing wires against the side wall of the vessel unless the patient has a significantly short life expectancy.

One of the biggest challenges when treating patients with any stent is maintaining patency. NICE anticoagulation guidance does not extend to use in patients with venous stents. At present, there is no evidence to suggest that one anticoagulant...
is superior in preventing post-procedure in-stent thrombosis; therefore, anticoagulation choice is largely personal. At St. Thomas’ Hospital, we treat a heterogeneous population, so our deep venous stenting program involves review of all patients by a multidisciplinary team with a view to optimizing anticoagulation on an individual basis.

ENDOVENOUS TREATMENT OF IVC THROMBOSIS

A number of studies have reported successful endovascular reconstruction of the IVC with acceptable short-term patency and significant early symptom relief. It is apparent that many cases of so-called IVC agenesis are in fact atretic and reconstructable provided the lesion can be crossed with a wire, which is achievable in the majority of cases.

There is little data to indicate the epidemiology of IVC atresia, but a history of long-line placement or neonatal umbilical vein catheterization is a frequent finding. The total absence of an IVC is a significant abnormality, and true agenesis is often associated with other significant abnormalities. It is therefore worth an attempt at reconstruction regardless of the appearance on imaging.

In acute presentation, many cases of IVC thrombosis are related to thromboses of the inflow vessels, and lysis may result in the collateral pathways being reopened. In these cases, it may be sufficient to bypass reconstruction and continue the patient on anticoagulation alone, reserving full reconstruction in case this treatment strategy is unsuccessful. In the case of IVC thrombosis precipitated by IVC filter placement, it is imperative to first try and retrieve the filter in conjunction with lytic therapy. If the filter is not retrievable, stenting across the filter is a well-described approach.

The IVC typically re-forms at the level of the renal veins or hepatic veins. Reconstruction needs to extend to the normal vessel, and these two points are the primary targets. If the suprarenal IVC is involved, it does not appear to be problematic to stent across the renal veins because the kidneys are usually drained by well-established collateral pathways.

Multiple techniques have been reported for reconstruction, and the one selected is largely dependent on the availability of stents. In the United States, the dominant techniques are based on use of the WALLSTENT endoprosthesis (Boston Scientific Corp.), whereas the increasing availability of dedicated nitinol venous stents outside of the United States has led to a dominance of double-barrel techniques.

Care should be taken during reconstruction when the underlying diagnosis is unrelated to thrombosis. In patients with extrinsic tumor compression or retroperitoneal fibrosis, the adjacent arteries may be compressed by the placement of venous stents. IVC rupture is also a known complication.

In the occurrence of IVC rupture, which is a more benign complication compared with SVC rupture, balloon tamponade can be used to allow bleeding to settle. If the defect is large and bleeding persists, then covered stents may be necessary.

As with endovenous treatments in the lower limb, success is predicated on the quality of the inflow vessels. Good inflow is closely linked to successful reconstruction, and the principle of stenting from “good to good” should be adhered to as closely as possible.

CONCLUSION

Reconstruction of the central veins is challenging, but the results of endovenous reconstruction are encouraging and improving with advances in techniques and equipment.

Much work is still required to create standard reporting criteria that allows comparison of techniques and patients.

KEY POINTS

- Inferior vena cava (IVC) and superior vena cava (SVC) reconstruction is technically feasible with endovenous techniques and demonstrates encouraging early results.
- The absence of a standardized classification system inhibits the ability to compare published data.
- Long-term results with IVC and SVC reconstruction are not yet established, but the emergence of dedicated venous stents and improved lytic techniques may continue to improve outcomes.
- There is a need for more robust data and longer-term follow-up to establish efficacy and durability of all techniques.

Conflict of Interest Disclosure:

Keywords:
thrombosis, central vein thrombosis, endovascular reconstruction, inferior vena cava, superior vena cava
REFERENCES


