JACC Cardiovasc Interv

STATE-OF-THE-ART REVIEW

Superior Vena Cava Syndrome

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Superior vena cava (SVC) syndrome comprises a constellation of clinical signs and symptoms caused by obstruction of blood flow through the SVC. The management of patients with life-threatening SVC syndrome is evolving from radiation therapy to endovascular therapy as the first-line treatment. There is a paucity of data and societal guidelines with regard to the management of SVC syndrome.

TABLE 2 Signs and Symptoms of SVC Syndrome

Signs and Symptoms	Incidence (%)
Facial edema	60-100
Nonpulsatile distended neck veins	27-86
Distended chest veins	38-67
Dyspnea and cough	23-70
Arm edema	14-75
Hoarseness and/or stridor	0-20
Syncope and/or headache	6-13
Confusion, obtundation	0-5

Signs and symptoms are presented in order of most common to least common. Adapted from Wilson et al. (2).

SVC = superior vena cava.

ENDOVASCULAR THERAPY

In the contemporary era, ET is used as the first-line therapy in majority of patients with SVC syndrome, particularly those

presenting with life-threatening symptoms such as cerebral or laryngeal edema or postural syncope.

TABLE 3 Grading of SVC Syndrome per Yu et al. (15)

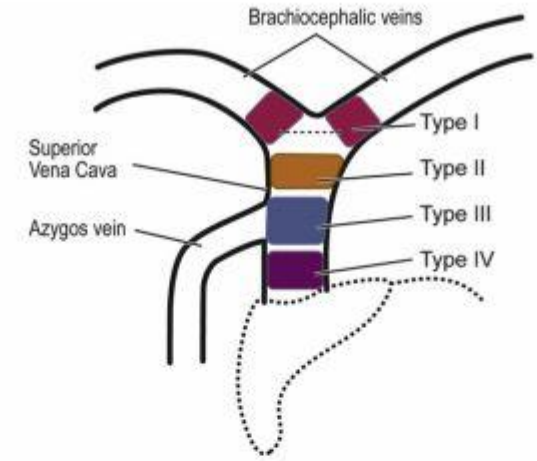
Grade	Finding(s)
0	Asymptomatic: SVC on imaging without symptoms
1	Mild: edema of head or neck
2	Moderate: edema in head or neck with functional impairment
3	Severe: mild or moderate cerebral edema/laryngeal edema, or diminished cardiac reserve
4	Life-threatening: significant cerebral edema, laryngeal edema, hemodynamic compromise
5	Fatal: death

SVC = superior vena cava.

Optimal treatment of pacemaker- or defibrillator related SVC syndrome is not well defined, as there is very little evidence to help guide clinicians.

The incidence of SVC syndrome after pacemaker or implantable cardioverter-defibrillator lead placement is a very rare adverse event, and routine anticoagulation as a preventive measure is not recommended even in high-risk patients, as the risks of bleeding outweighs the benefits.

FIGURE 3 Proposed Classification of SVC Obstruction Based on Location and Severity



Lesion location	Grade A Severity	Grade B severity	Grade C severity
Type I – Bilateral brachiocephalic vein occlusion with or without supra-azygos SVC	Moderate to severe (50-90%)	Pre-occlusive (>90%)	Totally occluded (100%)
Type II – Supra-Azygos SVC without brachiocephalic involvement	Moderate to severe (50-90%)	Pre-occlusive (>90%)	Totally occluded (100%)
Type III – Azygos SVC	Moderate to severe (50-90%)	Pre-occlusive (>90%)	Totally occluded (100%)
Type IV – Infra-Azygos SVC	Moderate to severe (50-90%)	Pre-occlusive (>90%)	Totally occluded (100%)

Proposed classification system that considers anatomic location and severity of SVC obstruction. When used in conjunction with clinical symptoms, it may be used to guide management and facilitate communication among clinicians. SVC = superior vena cava.

SVC SYNDROME WITH THROMBOSIS.

In some cases of SVC obstruction, there is superimposed thrombosis, likely related to stagnation of flow, a hypercoagulable state from underlying malignancy, or the presence of indwelling catheters. In such patients, the presentation is often acute and during intervention, the guide wire easily traverses the occluded segment. Thrombus removal with CDT or aspiration thrombectomy is recommended prior to

revascularization, in order to prevent pulmonary embolism and reduce the length of lesion to be treated. Thrombolysis or thrombectomy should be initiated within 2 to 5 days of symptom onset for treatment to be effective. After several days, the ability to achieve successful clot lysis drops significantly as the thrombus becomes organized. CDT has been shown to be safe and effective in patients both with and without cancer. If patients are at elevated risk of bleeding and have an absolute contraindication to thrombolytic agents, then mechanical thrombectomy may be preferred.

ENDOVASCULAR TECHNIQUES

Most of these procedures are performed in a supine position in the endovascular suite with local anesthesia and conscious sedation. Some patients may need to have the head elevated or need general

Anesthesia with mechanical ventilation depending on if symptoms worsen by lying flat or the presence of airway edema, respectively.

In patients with non-occlusive lesions, a single access site is often sufficient, although dual access can provide more options for imaging and better support for device delivery. Many operators prefer the femoral approach for convenience. In patients with total SVC occlusion, additional access is advised, for successful

recanalization. Options for access cephalad to the SVC include the basilic, brachial, axillary, or internal jugular veins. Many operators will first attempt crossing with a hydrophilic wire (e.g., glide wire, angled or straight, stiff, or floppy), supported by diagnostic catheter. Once the lesion is crossed successfully, pre-dilation with an undersized balloon (2 to 4 mm) of the stenosis is recommended. This is followed with sequential dilation with increasing balloon diameters, assessing the response of the vessel and the clinical reaction of the patient (e.g., pain) with each dilation. Pre-dilation facilitates stent deployment and expansion, but oversizing the balloons to >16 mm has been associated with increased risk of SVC rupture, dysrhythmias, pericardial tamponade, or cardiac arrest.

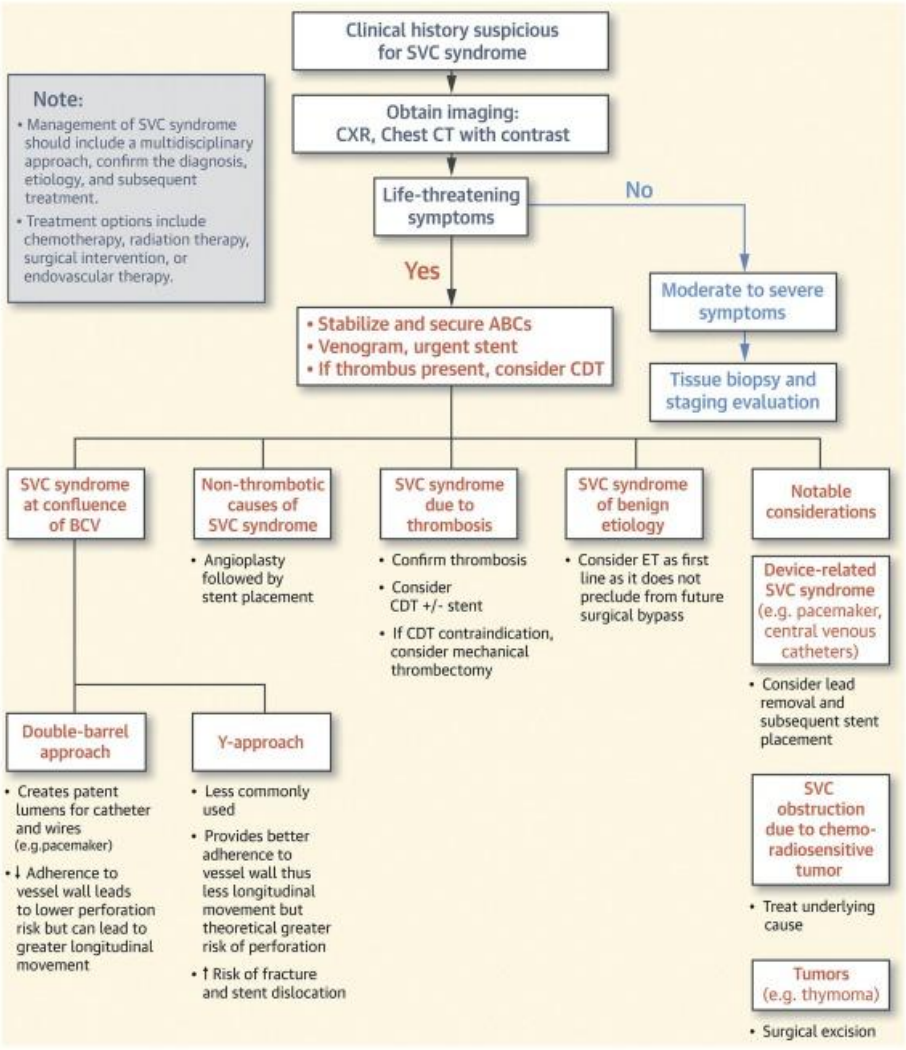
TYPE OF STENTS:

TABLE 5 Venous Stents, Types, Descriptions, Benefits, and Drawbacks

Type of Stent	Description	Benefits	Drawbacks
Wallstent endoprosthesis	Elgiloy braided construction self-expanding Available in large diameters	Well studied and widely used Provides stent flexibility and efficacious in long stenosis	Flexible and undergoes shortening after deployment leading to decreased deployment accuracy Prone to migration Poor radial strength
Palmaz stent	Balloon-expandable stent	High radial force useful in lesions with recoil Allows staged dilatation of stenosis to larger diameter Lower propensity for stent migration	Relatively incompressible, which can lead to stent fracture and reocclusion
Gianturco Z-stent	25–30 mm in diameter	Preferable for isolated SVC stenosis Small circumferential hooks prevent migration Allows precise positioning	Rigid, making placement into tortuous vessels difficult Open-cell design and low contact with vessel wall may not inhibit tumor ingrowth as effectively as other stents do Given diameter, requires wide bore sheath (up to 16-F) to deploy
Zilver Vena venous stent	Open-cell design Available in 14–16 mm diameters and 60–140 mm lengths	Created for venous system Offers flexibility and minimal foreshortening	Limited data
Venovo stent	Open-cell design 8-F to 10-F platform and comes in 10–20 mm diameters and 40–160 mm lengths	Created for venous system Evidence has shown high patency rates in iliofemoral venous obstruction	Has not been widely used for SVC syndrome and data lacking
VICI stent	Available in 12–16 mm diameters and 60–140 mm lengths	Very good radial force	Limited data
Viatorr stent	Endoprosthesis designed for TIPS to treat portal hypertension Longest stent is 10 cm	Combination of bare stent and covered stent Conformable to tortuous anatomy	Not widely used in SVC syndrome Long-term patency of bare section of stent outside of TIPS has not been studied
Viabahn VBX stent-graft	Balloon-expandable covered endoprosthesis Composed of independent 316 L stainless steel ring elements that are connected via a heparinized fluoropolymer graft material	Minimum slippage Only covered stent lacking longitudinal struts for ring linkage allowing maximal flexibility while maintaining excellent radial strength	Food and Drug Administration approved for iliac artery stenosis Limited data for SVC syndrome

Practical approach for treatment of SVC syndrome:

CENTRAL ILLUSTRATION: Proposed Management Algorithm of SVC Syndrome Based on Severity of Symptoms and Common Etiologies



Azizi, A.H. et al. J Am Coll Cardiol Intv. 2020;13(24):2896-910.