

# BMJ Best Practice

## Superior vena cava syndrome

Straight to the point of care



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## Summary

Superior vena cava (SVC) syndrome is a clinical condition that occurs due to obstruction of the SVC.

The most common etiology is malignancy; however, there has been an increase in benign causes due to more frequent use of intravascular devices.

Although rarely fatal, it may sometimes present as life-threatening upper airway obstruction.

A high index of suspicion is required to make the diagnosis in many cases.

Treatment and prognosis depend on underlying etiology.

## Definition

Superior vena cava (SVC) syndrome is a clinical condition that occurs as a result of obstruction of the SVC, leading to interrupted venous return from the head, thorax, and upper extremities to the right atrium. The increased venous pressure results in edema of the head, neck, and arms, often with cyanosis, plethora, and distended subcutaneous vessels.<sup>[1] [2]</sup> It can be caused by either intraluminal obstruction of the SVC or extrinsic compression.

## Epidemiology

Superior vena cava (SVC) syndrome occurs in approximately 15,000 people in the US every year.[1] Malignancy causes 65% of cases, lung cancer and non-Hodgkin lymphoma being the most common. Malignant causes of SVC syndrome are more frequent in middle-aged to older adult males, while benign causes are equally distributed across both sexes and more commonly seen in the younger population. Infectious causes (especially syphilitic aortic aneurysm and tuberculosis) accounted for the majority of cases 50 years ago but are now rare, especially in developed countries.[7] Malignant causes accounted for >90% of cases around 25 years ago, but there has been an increase in benign causes of SVC syndrome, reflecting increased use of intravascular devices such as catheters, pacemakers, implantable cardioverter-defibrillators, and cardiac resynchronization therapy.[8]

## Etiology

A total of 65% to 70% of cases are due to malignancy.[1] [2] Lung cancer is the most common etiology, with non-small cell lung cancer accounting for 50% of cases of malignant superior vena cava (SVC) syndrome and small cell lung cancer for 25% cases.[9] [10] [11] Overall, 2% to 5% of all patients with lung cancer go on to develop SVC syndrome, while 10% to 20% of patients with small cell lung cancer develop the condition. This is most likely related to the central growth of these tumors.[12] Most patients with lung cancer-associated SVC syndrome have right-sided lesions (80%). Lymphoma is the second most common cause, accounting for 12% of cases of malignant SVC syndrome; diffuse large cell lymphoma is the most common type (two-thirds), followed by lymphoblastic lymphoma (one third). Of patients with primary mediastinal B-cell lymphoma with sclerosis (a rare subtype of non-Hodgkin lymphoma), the prevalence of SVC syndrome is high, with one study reporting 57%.[13] Although Hodgkin lymphoma often involves the mediastinum, SVC syndrome is rare.[14] Thymoma (2%) and germ cell tumors (3%) are other primary mediastinal malignancies that occasionally cause SVC syndrome.[15]

The most common metastatic disease that causes SVC syndrome is breast cancer, accounting for 11% of cases.[6] Other metastatic tumors that cause SVC syndrome include colon cancer, esophageal cancer, Kaposi sarcoma, and fibrous mesothelioma.

Benign causes of SVC syndrome are less frequent (30%-35%).[1] [2] They include iatrogenic causes associated with SVC thrombosis (e.g., central venous catheters, pacemaker and implantable cardioverter-defibrillator leads), mediastinal fibrosis caused by radiation therapy or infections (e.g., histoplasmosis, tuberculosis, aspergillosis, blastomycosis, or nocardiosis), collagen-vascular diseases like sarcoidosis or Behcet syndrome, and, rarely, aortic arch aneurysm, large substernal goiter, mediastinal hematoma as a result of trauma, and bicaval anastomotic stenosis following cardiac transplantation.[1] [11] [16] Complications of pacemaker lead placement, such as venous thrombosis and stenosis, occur in up to 30% of patients, but only a few patients become symptomatic; however, the presence of multiple leads, retention of severed lead(s), and previous lead infection may increase risk of SVC syndrome.

In children, SVC syndrome is most often caused by non-Hodgkin lymphoma. The compression of the SVC may be associated with compression of the trachea, which is narrow, flexible, and soft relative to that of an adult. This may result in airway obstruction in children.

## Pathophysiology

The superior vena cava (SVC) extends from the junction of the right and left brachiocephalic veins to the right atrium. It drains venous blood from the head, neck, upper extremities, and upper thorax to the right atrium. It is located in the middle mediastinum and is surrounded by structures including the trachea, right bronchus, aorta, pulmonary artery, and the perihilar and paratracheal lymph nodes.

The thin-walled SVC can be obstructed by intraluminal, mural, or extrinsic factors. The extent and rapidity of obstruction correlates with elevation of venous pressure and symptomatic presentation. Slowly progressive obstruction leads to recruitment of collateral circulation via the azygos and internal mammary venous system (which takes several weeks).[17] With obstruction of the SVC, the cervical venous pressure is usually increased to 20 to 40 mmHg (normal range 2-8 mmHg).[18] When obstruction occurs abruptly, SVC syndrome can constitute a medical emergency.

An obstructed SVC initiates collateral venous return to the heart from the upper half of the body through four principal pathways. The most important pathway is the azygos venous system, which includes the azygos vein, the hemiazygos vein, and the connecting intercostal veins. The second pathway is the internal mammary venous system plus tributaries and secondary communications to the superior and inferior epigastric veins. The long thoracic venous system, with its connections to the femoral veins and vertebral veins, provides the third and fourth collateral routes, respectively.[1]

## Classification

### Classification based on location of superior vena cava (SVC) obstruction

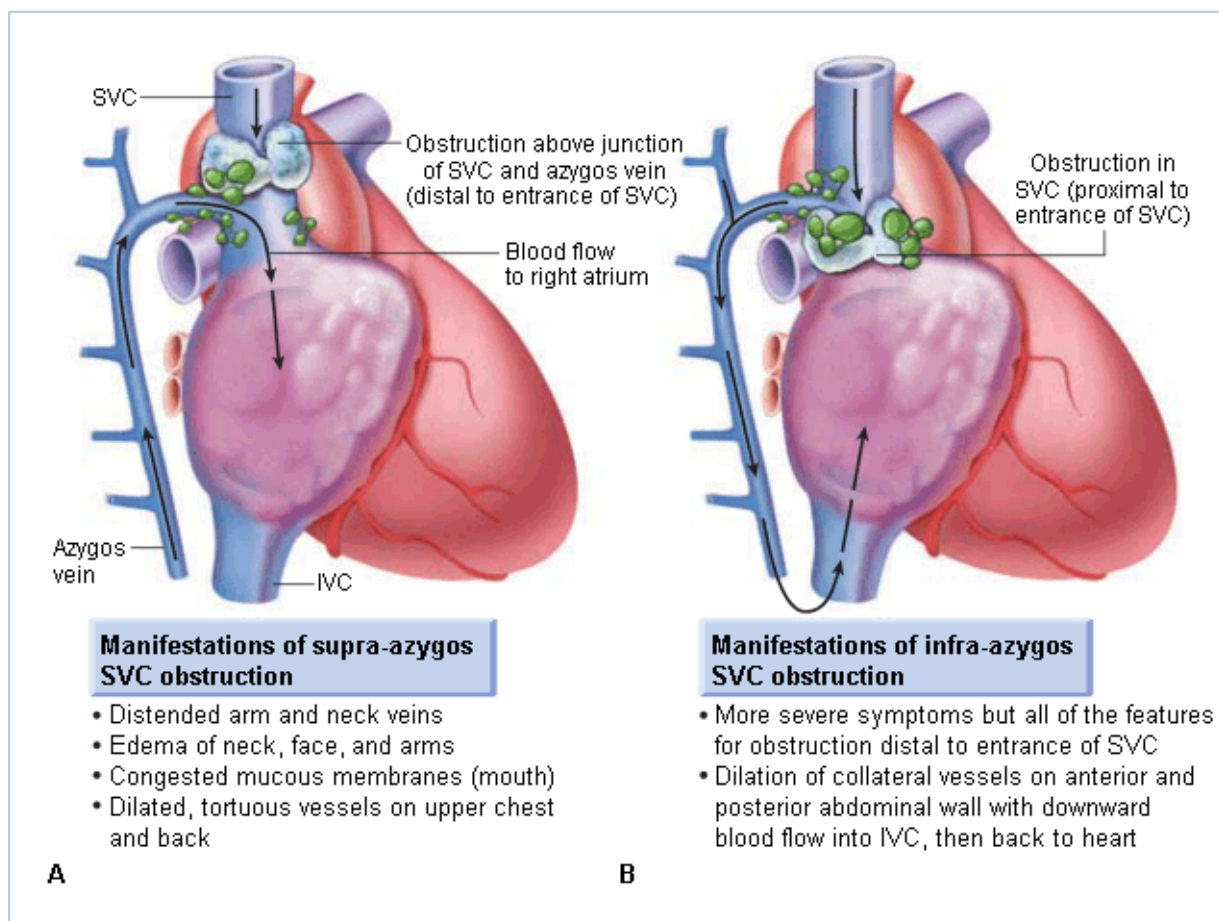
Preazygos or supra-azygos

- Obstruction of blood return above the entrance of azygos vein into the SVC, resulting in venous distension and edema of the face, neck, and upper extremities.[3] [4]

Postazygos or infra-azygos

- Obstruction below the entrance of azygos vein into the SVC results in retrograde flow through the azygos via collaterals to the inferior vena cava, resulting in not only the symptoms and signs of preazygos disease, but also dilation of the veins over the abdomen.
- This is usually more severe and poorly tolerated than preazygos obstruction.





*Supra- and infra-azygos obstruction leading to superior vena cava (SVC) syndrome. IVC: inferior vena cava*

*Reproduced with permission from Braunwald's Heart Disease, 8th ed (2008)*

## Classification based on etiology of obstruction

- Luminal obstruction (e.g., pacemaker leads or catheter-related thrombosis).
- Extrinsic compression (e.g., malignancy, fibrosing mediastinitis due to infection/radiation, aortic arch aneurysm, hematoma, or goiter).

## Case history

### Case history #1

A 65-year-old man with a 40-year history of chronic smoking, hypertension, and chronic obstructive pulmonary disease presents with anorexia and weight loss for the past 6 months. He had been complaining of increased dyspnea with exertion and orthopnea, and has noticed bilateral arm swelling and facial plethora for the past 3 weeks. At the time of admission, his face and upper extremities were edematous, and there were engorged veins in his neck and upper extremity.

### Case history #2

A 70-year-old woman with a history of ischemic cardiomyopathy, left ventricular ejection fraction of 25%, and prior history of cardiac resynchronization therapy 3 years ago presents with slowly progressive swelling of the face and both arms, as well as prominent veins in the neck and upper extremities. She

gives a history of excessive bleeding from the site of venepuncture in the antecubital region after blood draws in the last few months.

## Other presentations

Atypical presentations may include presentation to the emergency department with sudden-onset dyspnea due to laryngeal edema that can be acutely fatal due to upper airway compromise.<sup>[5]</sup> In addition, cyanosis, papilledema, cerebral edema, mental changes, stupor, and even coma have been described with severe obstruction.<sup>[1] [6]</sup>

## Approach

The diagnosis of superior vena cava (SVC) syndrome is usually clinical and requires a high degree of suspicion; thus, a good history and physical examination is important. Chest CT/MRI is the initial test of choice required to confirm the diagnosis and to evaluate for underlying etiology. Obtaining tissue for histopathology may be required in cases of suspected malignancy or infection.

### History

History may also be notable for smoking, multiple pacemaker leads, or central venous catheters. Previous history of radiation exposure should be noted, as excess radiation to the mediastinum can lead to fibrosis causing SVC obstruction.

Early in the course, partial SVC obstruction may be asymptomatic or associated with subtle symptoms. With progressive obstruction, the classic symptoms and signs become more obvious. The most common symptoms are facial swelling, dyspnea, cough, arm swelling, and facial plethora.<sup>[1] [2]</sup> Sudden-onset dyspnea due to laryngeal edema can be fatal as a result of upper airway compromise.<sup>[5]</sup> Other symptoms include headache, chest pain, blurred vision, hoarseness of voice, and stridor. Symptoms are usually worsened by bending forward or lying down.

History of associated anorexia, weight loss, cough, dyspnea, and hemoptysis may suggest lung malignancy (or possibly chronic infection). Fever, skin rash, or arthralgias may be indicative of underlying collagen-vascular diseases.

### Physical examination

Examination usually reveals engorged veins of the neck and upper chest wall, facial edema, and upper-extremity edema. Prominent collateral veins covering the anterior chest wall may be visible. The maneuver of bending forward usually worsens venous engorgement and is a helpful clinical sign. Laryngeal edema, cyanosis, papilledema, mental changes, stupor, and even coma have been described with severe obstruction.<sup>[1] [6]</sup> When lymphadenopathy is present outside the chest, lymphoma should be considered a possibility.

### Investigations

Chest x-ray may be performed as an initial test and sometimes reveals a widened mediastinum or mass lesion in the lung, but the most important radiologic investigation when diagnosis is clinically suspected is CT of the chest (with intravenous contrast).<sup>[1] [2]</sup> It establishes the diagnosis of SVC obstruction and shows the exact location, severity, and associated pathology (e.g., malignancy or intravascular thrombosis). MRI may also be useful but does not have any distinct advantages over CT, except in patients with contrast allergy or renal failure, as it avoids iodinated contrast.<sup>[1] [2]</sup>

Ultrasound of the upper extremities is a useful noninvasive screening test and helps in identification of venous thrombosis or obstruction.<sup>[2] [19]</sup> Presence of monophasic flow in the SVC or loss of respiratory variation on Doppler ultrasound can suggest SVC obstruction.

Bilateral upper-extremity venography can accurately delineate the site and extent of SVC obstruction and collateral pathways, but does not provide information about lung and mediastinal pathology. Venography is usually not required for diagnosis, but may be helpful to plan stent placement or surgery.<sup>[1] [2]</sup>



## Other investigations

Obtaining a tissue diagnosis is important to confirm the presence of malignancy. A biopsy from supraclavicular or other cervical lymph nodes may obviate the need for invasive procedures like mediastinoscopy, and thus careful examination for cervical lymphadenopathy should be performed. For diagnosis of malignancy, bronchoscopy has a diagnostic yield of 50% to 70%, transthoracic needle-aspiration biopsy has a yield of approximately 75%, and mediastinoscopy or mediastinotomy has a diagnostic yield of >90%.<sup>[1]</sup>

Sputum examination for culture, acid-fast staining, and cytology is helpful in diagnosis of cases with tuberculosis, fungal infections (e.g., aspergillosis, blastomycosis, histoplasmosis, nocardiosis), or endobronchial malignancy. Thoracentesis with cytologic analysis should be strongly considered whenever pleural effusion is present.

Erythrocyte sedimentation rate or C-reactive protein may be elevated in patients with infection or immunologic disorders.

## History and exam

### Key diagnostic factors

#### history of smoking, pacemaker, or central venous catheter (common)

- Lung cancer is the most common overall cause of superior vena cava syndrome, while multiple pacemaker leads and central venous catheters are becoming increasingly frequent benign causes of the condition.

#### localized edema of the face and upper extremities (common)

- Present in 80% of cases.<sup>[9]</sup>
- If edema is localized to upper extremities and face, obstruction of the superior vena cava should be considered.

#### dyspnea (common)

- Present in 60% of cases.<sup>[20]</sup>
- Usually made worse by bending forward or lying down (orthopnea).
- May suggest lung malignancy or chronic infection.

#### facial plethora (common)

- Due to venous engorgement and edema.

#### cough (common)

- Present in 54% of cases.<sup>[10]</sup>
- Can be related to underlying etiology or laryngeal edema.

#### distended neck veins (common)

- Seen in 63% of cases and due to increased venous pressure.<sup>[1]</sup>
- Bending forward usually worsens venous engorgement and is a helpful clinical sign.

**distended chest veins (common)**

- Seen in 53% of cases and due to increased venous pressure.[1]
- Prominent collateral veins covering the anterior chest wall may be visible.
- Bending forward usually worsens venous engorgement and is a helpful clinical sign.

**hoarseness of voice (common)**

- Present in 17% of cases.[6]
- Can be related to underlying etiology or laryngeal edema.

**lymphadenopathy (common)**

- Lymphoma is a possibility if lymphadenopathy is outside of the chest.

**blurred vision (uncommon)**

- Present in 2% of cases.[6]

**stridor (uncommon)**

- Present in 4% of cases.[6]
- Related to laryngeal edema or direct compression.

**confusion/stupor (uncommon)**

- Present in 4% of cases and due to cerebral edema.
- Has been described with severe obstruction.[1] [6]

**Other diagnostic factors****anorexia (common)**

- May suggest lung malignancy or chronic infection.

**weight loss (common)**

- May suggest lung malignancy or chronic infection.

**hemoptysis (common)**

- May suggest lung malignancy or chronic infection.

**headache (uncommon)**

- Present in 9% of cases.[6]
- Due to increased cerebral venous pressure.

**chest pain (uncommon)**

- Usually pleuritic; related to pleural involvement from malignancy, infection, or autoimmune diseases.

**mental changes (uncommon)**

- Has been described with severe obstruction.[1] [6]

**fever (uncommon)**

- May be indicative of collagen-vascular disease.

**skin rash (uncommon)**

- May be indicative of collagen-vascular disease.

**arthralgia (uncommon)**

- May be indicative of collagen-vascular disease.

**laryngeal edema (uncommon)**

- Has been described with severe obstruction.[\[1\]](#) [\[6\]](#)

**cyanosis (uncommon)**

- Has been described with severe obstruction.[\[1\]](#) [\[6\]](#)

**papilledema (uncommon)**

- Has been described with severe obstruction.[\[1\]](#) [\[6\]](#)

**coma (uncommon)**

- Has been described with severe obstruction.[\[1\]](#) [\[6\]](#)

## Risk factors

**Strong****smoking**

- Strong relationship to lung cancer, the most common overall cause of superior vena cava syndrome.

**multiple pacemaker leads**

- Becoming an increasingly frequent benign cause of superior vena cava syndrome.

**central venous catheters/ports**

- Becoming an increasingly frequent benign cause of superior vena cava syndrome.[\[7\]](#)

**Weak****age >50 years**

- Lung malignancy should be considered as the most likely etiology in patients >50 years of age.

**radiation**

- Excess radiation to the mediastinum can lead to fibrosis causing superior vena cava obstruction.

## Tests

### 1st test to order

Test	Result
<b>chest x-ray</b> <ul style="list-style-type: none"> <li>Ordered when superior vena cava syndrome is clinically suspected, especially with a history of pulmonary symptoms.</li> </ul>	<b>widened mediastinum or mass lesion in the lung</b>
<b>chest CT</b> <ul style="list-style-type: none"> <li>Most useful imaging test as it helps to establish diagnosis.<sup>[1] [2]</sup></li> <li>Undertaken with intravenous contrast.</li> <li>Ordered when there is clinical suspicion of superior vena cava syndrome.</li> <li>Helpful in obtaining a tissue diagnosis by CT-guided biopsy.</li> </ul>	<b>full or partial obstruction; development of collateral vessels; shows location, severity, and associated pathology (e.g., malignancy or intravascular thrombosis)</b>
<b>chest MRI</b> <ul style="list-style-type: none"> <li>Useful in patients with a history of contrast allergy or those at risk of contrast-induced worsening of renal function.<sup>[1] [2]</sup></li> <li>Caution advised in use of gadolinium in renal insufficiency due to risk of nephrogenic fibrosing dermopathy.</li> <li>Contraindicated in patients with pacemakers and defibrillators.</li> </ul>	<b>full or partial obstruction; development of collateral vessels; shows location, severity, and associated pathology (e.g., malignancy or intravascular thrombosis)</b>
<b>ultrasound of upper extremities</b> <ul style="list-style-type: none"> <li>Useful noninvasive screening test.</li> <li>Helps in identification of venous thrombosis or obstruction.<sup>[2] [19]</sup></li> <li>Presence of monophasic flow in the superior vena cava (SVC) or loss of respiratory variation on Doppler ultrasound can suggest SVC obstruction.</li> </ul>	<b>dilated SVC; presence of thrombus; monophasic flow; loss of respiratory variation</b>

## Other tests to consider

Test	Result
<b>venography</b> <ul style="list-style-type: none"> <li>Invasive test, usually performed by venous catheterization through the femoral vein and injection of contrast dye in the superior vena cava (SVC).</li> <li>Does not provide information about lung or mediastinal pathology.[1]</li> <li>Not usually required for diagnosis, but may be useful for planning endoscopic interventions or before surgery.[1] [2]</li> </ul>	<b>defines site and extent of SVC obstruction and collateral pathways</b>
<b>biopsy</b> <ul style="list-style-type: none"> <li>Obtaining tissue diagnosis is important to confirm presence of malignancy.</li> <li>Bronchoscopy has a diagnostic yield of 50% to 70%, transthoracic needle-aspiration biopsy has a yield of approximately 75%, and mediastinoscopy or mediastinotomy has a diagnostic yield of &gt;90%.[1]</li> <li>Biopsy from supraclavicular or other cervical lymph node may obviate the need for invasive procedures like mediastinoscopy and, thus, careful examination for cervical lymphadenopathy should be performed.</li> </ul>	<b>specimen for pathologic diagnosis</b>
<b>sputum cytology</b> <ul style="list-style-type: none"> <li>Simple, noninvasive method to detect lung malignancy. More likely to be positive with central lesions than with peripheral lesions.</li> <li>Thoracentesis with cytologic analysis should be strongly considered when pleural effusion is present.</li> </ul>	<b>malignant cells in sputum</b>
<b>thoracentesis</b> <ul style="list-style-type: none"> <li>Thoracentesis involves placing a needle between the ribs and into the chest to sample fluid that has accumulated in the pleural space.</li> <li>Thoracentesis with cytologic analysis should be strongly considered whenever pleural effusion is present.</li> </ul>	<b>malignant cells in pleural fluid</b>
<b>sputum culture</b> <ul style="list-style-type: none"> <li>Sputum examination for culture is helpful in diagnosis of cases with tuberculosis, or bacterial or fungal infections (e.g., aspergillosis, blastomycosis, histoplasmosis, nocardiosis).</li> </ul>	<b>growth of specific organisms</b>
<b>erythrocyte sedimentation rate</b> <ul style="list-style-type: none"> <li>May be present in patients with infection or immunologic disorders.</li> </ul>	<b>elevated</b>
<b>C-reactive protein</b> <ul style="list-style-type: none"> <li>May be present in patients with infection or immunologic disorders.</li> </ul>	<b>elevated</b>

## Differentials

Condition	Differentiating signs / symptoms	Differentiating tests
<b>Cardiac tamponade</b>	<ul style="list-style-type: none"> <li>• Absence of facial and upper-extremity edema.</li> <li>• Variation of jugular venous pressure with respiration (prominent x-descent).</li> <li>• Pulsus paradoxus present.</li> </ul>	<ul style="list-style-type: none"> <li>• Pericardial effusion is seen on CT chest.</li> <li>• Echocardiography shows bouncing septum, marked respiratory variation in the early left ventricular filling velocity (&gt;25%), and right ventricular diastolic collapse.</li> </ul>
<b>Constrictive pericarditis</b>	<ul style="list-style-type: none"> <li>• Elevated jugular venous pressure (JVP) with prominent negative descents (x- and y-descent).</li> <li>• Presence of Kussmaul sign (increase in JVP with inspiration).</li> </ul>	<ul style="list-style-type: none"> <li>• Echocardiography may show thickened pericardium and marked respiratory variation in the early left ventricular filling velocity (&gt;25%).</li> <li>• MRI is the investigation of choice, as it shows pericardial thickening and ventricular interdependence.</li> <li>• Cardiac catheterization shows discordance of left and right ventricular pressures with respiration, which has high specificity for diagnosis.</li> </ul>
<b>Acute COPD exacerbation</b>	<ul style="list-style-type: none"> <li>• Extensive bilateral expiratory wheezing, hypoxia, and hypercarbia.</li> </ul>	<ul style="list-style-type: none"> <li>• Peak flow, spirometry, and bronchodilator response help in differentiating.</li> <li>• Presence of obstructive defect on pulmonary function testing is seen.</li> </ul>
<b>Right-sided heart failure</b>	<ul style="list-style-type: none"> <li>• Preserved respiratory variation in jugular venous pressure, prominent negative descents, and sometimes increased v wave due to tricuspid regurgitation.</li> </ul>	<ul style="list-style-type: none"> <li>• Echocardiography will show right ventricular dysfunction and dilated inferior vena cava with lack of inspiratory collapse.</li> </ul>
<b>Pulmonary embolism</b>	<ul style="list-style-type: none"> <li>• Upper-extremity edema is usually absent.</li> </ul>	<ul style="list-style-type: none"> <li>• CT chest with contrast will show presence of thrombus inside the pulmonary artery.</li> </ul>
<b>Cardiac tumor</b>	<ul style="list-style-type: none"> <li>• Upper-extremity edema is usually absent.</li> </ul>	<ul style="list-style-type: none"> <li>• Echocardiography or cardiac MRI will show presence of a mass, usually inside the right side of the heart.</li> </ul>



## Approach

Once diagnosis has been established, a malignant or nonmalignant cause of superior vena cava (SVC) syndrome must be determined, as treatment options differ. Treatment involves relieving the symptoms of obstruction and treating the underlying etiology. There have been no large randomized trials to compare various treatment options, and most data are from case series and expert opinion. Treatment of SVC syndrome is interdisciplinary and may involve oncology, respiratory, surgery, radiology, vascular, and endovascular specialists.[2]

### Acute airway obstruction (without tissue diagnosis)

Presentation with airway obstruction is serious, although rare in current clinical practice. First-line treatment consists of securing the airway and relief of obstructive symptoms. This can be achieved with either a combination of corticosteroids and radiation therapy, or percutaneous stenting.[2] Urgent treatment with radiation therapy and corticosteroids should be used only for life-threatening situations. It should be deferred otherwise due to interference with subsequent histopathologic diagnosis. Stenting is increasingly used because the stent can be placed before a tissue diagnosis is available. It is a useful procedure for patients with severe symptoms such as respiratory distress that require urgent intervention.[21] [22] Meta-analyses have demonstrated that endovascular therapy with stenting has high technical and clinical success rates.[23] [24] [25]

In the absence of a need for urgent intervention, the management should focus initially on establishing the correct diagnosis.

### Malignant obstructions

Malignant causes require further treatment with appropriate chemotherapy, radiation, and/or surgery. Most malignant tumors causing SVC syndrome are sensitive to radiation therapy. Chemotherapy is an effective option for treatment of lung cancer, lymphomas, and germ cell tumors.[26] Thymomas that are resistant to chemotherapy and radiation may require surgical resection and SVC reconstruction.[27] Selection of therapy will depend on the type of malignancy, staging, and histopathology. See Small cell lung cancer , Non-small cell lung cancer , Non-Hodgkin lymphoma , Thymic tumor .

Endovascular stenting is performed to achieve more rapid improvement in symptoms and has fewer side effects compared with radiation therapy.[23] [24] [25]

Second line treatment is palliative therapy. This includes palliative radiation therapy, chemotherapy or corticosteroids (for lymphomas and thymomas), endovascular stents, or rarely bypass surgery.[1] [2] In rare cases, surgical decompression can be performed. Thrombolysis with indwelling catheters has also been described in small studies.[28] Supportive treatment consists of diuretics, low-salt diet, avoidance of upper-extremity lines, head elevation, and oxygen.

### Benign obstructions

Benign causes can be managed with percutaneous stenting, intravascular thrombolysis, bypass grafting, anticoagulation, or treatment of underlying infectious etiology.

Underlying infection (e.g., aspergillosis, blastomycosis, histoplasmosis, nocardiosis) should be treated according to local sensitivities. Endovascular stents and more rarely bypass surgery may be required if SVC obstruction persists after treatment of infection.

Catheter(s) should be removed and local thrombolysis and/or short-course anticoagulation should be considered in patients with thrombosis due to central venous catheter(s).<sup>[29]</sup>

Percutaneous balloon dilatation/stenting is preferred in patients with pacemaker and implantable cardioverter-defibrillator lead-related venous occlusion. Lead explantation may carry a high risk of mortality.<sup>[30]</sup> Bypass surgery may be an option. Infection of the leads should always be considered as a possibility and evaluated with blood cultures and transesophageal echocardiogram. Anticoagulation with warfarin should be considered. Data regarding post-procedural anticoagulation are lacking, and practices vary.<sup>[2]</sup>

## Treatment algorithm overview

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: [see disclaimer](#)

Acute ( summary )		
acute airway obstruction (without tissue diagnosis)		
	1st	secure airway + radiation therapy + corticosteroids
	1st	secure airway + percutaneous endovascular stenting
Ongoing ( summary )		
malignant etiology		
	1st	treatment of malignancy
	plus	percutaneous endovascular stenting
	2nd	palliative therapy
infectious etiology		
	1st	treatment of infection
	2nd	palliative therapy
iatrogenic etiology		
■ thrombosis due to central venous catheter(s)	1st	catheter removal + thrombolysis and/or anticoagulation
■ pacemaker and implantable cardioverter-defibrillator lead-related venous occlusion	1st	percutaneous balloon dilatation/stenting with or without lead removal

# Treatment algorithm

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: [see disclaimer](#)

## Acute

### acute airway obstruction (without tissue diagnosis)

#### 1st secure airway + radiation therapy + corticosteroids

##### Primary options

» [dexamethasone sodium phosphate](#): 10 mg intravenous bolus initially, followed by 4 mg every 6 hours

» Airway should be secured by intubation or surgically first.

» Urgent treatment with radiation therapy and corticosteroids should be used only for life-threatening situations. It should be deferred otherwise, due to interference with subsequent histopathologic diagnosis.

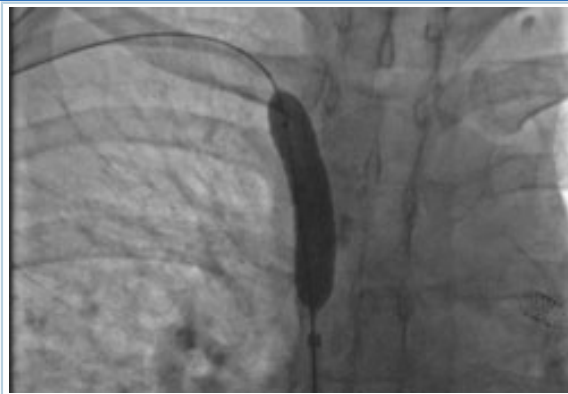
» Dose of corticosteroids should be limited and the dose decreased after 1 to 2 days of treatment or following symptomatic improvement.

#### 1st secure airway + percutaneous endovascular stenting

» Airway should be secured by intubation or surgically first.

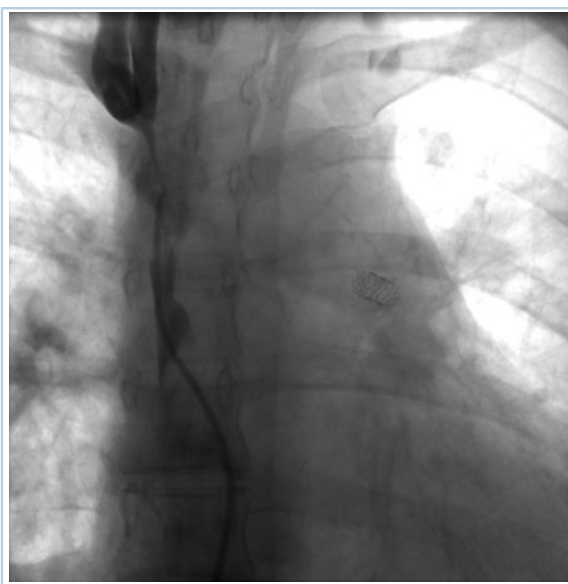
» Becoming increasingly used, because the stent can be placed before a tissue diagnosis is available. It is a useful procedure for patients with severe symptoms such as respiratory distress that require urgent intervention.<sup>[21]</sup><sup>[22]</sup> Meta-analyses have demonstrated that endovascular therapy with stenting has high technical and clinical success rates.<sup>[23]</sup> <sup>[24]</sup> <sup>[25]</sup>

» Done percutaneously by obtaining access usually through the femoral vein. Performed under conscious sedation. Fluoroscopic guidance and iodinated contrast are used. Most operators use heparin during the procedure.

**Acute**

*Postdilatation of the superior vena cava stent*

*Image obtained from cardiac catheterization laboratory at University of Missouri, Columbia; used with permission*

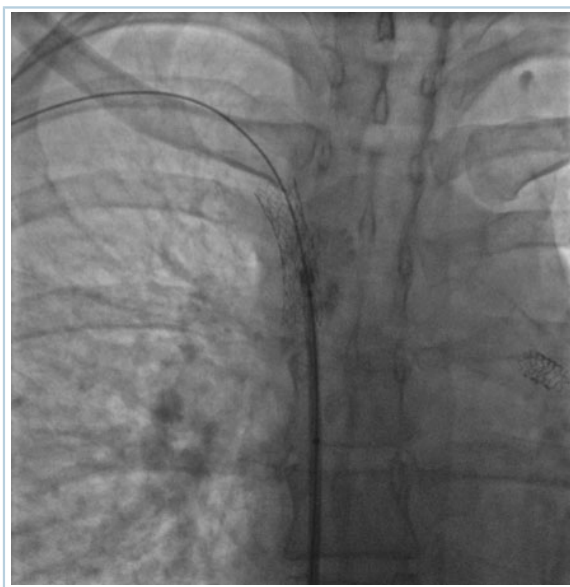


*Venography showing superior vena cava stenosis. Stent placement in the left pulmonary artery is seen*

*Image obtained from cardiac catheterization laboratory at University of Missouri, Columbia; used with permission*

**Acute**

*Percutaneous balloon angioplasty of the stenotic lesion in superior vena cava  
Image obtained from cardiac catheterization laboratory at University of Missouri, Columbia; used with permission*



*Stent deployment in the superior vena cava  
Image obtained from cardiac catheterization laboratory at University of Missouri, Columbia; used with permission*

- » Self-expanding or balloon-expandable stents may be used (usually bare metal stents).
- » Complications of percutaneous stenting are in the range of 3% to 7% and include volume overload due to sudden increase in preload, stent thrombosis, pulmonary embolus, stent migration, hematoma at the insertion site,

## Acute

infection, bleeding, and, very rarely, perforation or death.[\[22\]](#)

» Patency rate is around 80% to 94%, and 20% of patients may require repeat stenting.

» Bleeding risk is 1% to 14%, due to anticoagulation with aspirin, clopidogrel, and/or warfarin, which may be used following stent placement to prevent thrombosis.[\[21\]](#) [\[31\]](#)

» Data regarding post-procedural anticoagulation are lacking, and practices vary.[\[2\]](#)



## Ongoing

## malignant etiology

**1st treatment of malignancy**

- » Most malignant tumors causing superior vena cava (SVC) syndrome are sensitive to radiation therapy.
- » Chemotherapy is an effective option for treatment of lung cancer, lymphomas, and germ cell tumors.[26]
- » Thymomas that are resistant to chemotherapy and radiation may require surgical resection and SVC reconstruction (operative mortality rate of 5% and patency rate of 80% to 90%).[27]
- » Selection of therapy will depend on the type of malignancy, staging and histopathology. See Small cell lung cancer , Non-small cell lung cancer , Non-Hodgkin lymphoma , Thymic tumor .

**plus percutaneous endovascular stenting**

Treatment recommended for ALL patients in selected patient group

- » Endovascular stenting is performed to achieve more rapid improvement in symptoms and has fewer side effects compared with radiation therapy.[23] [24] [25]
- » Undertaken percutaneously by obtaining access (usually) through the femoral vein. Performed under conscious sedation. Fluoroscopic guidance and iodinated contrast are used and most operators use heparin during the procedure.



*Postdilatation of the superior vena cava stent  
Image obtained from cardiac catheterization  
laboratory at University of Missouri,  
Columbia;#used with permission*

## Ongoing

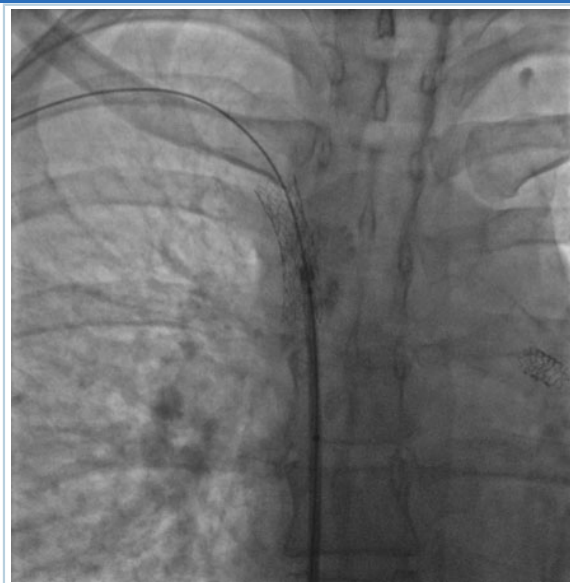


*Venography showing superior vena cava stenosis. Stent placement in the left pulmonary artery is seen  
Image obtained from cardiac catheterization laboratory at University of Missouri, Columbia; used with permission*



*Percutaneous balloon angioplasty of the stenotic lesion in superior vena cava  
Image obtained from cardiac catheterization laboratory at University of Missouri, Columbia; used with permission*

## Ongoing



*Stent deployment in the superior vena cava  
Image obtained from cardiac catheterization  
laboratory at University of Missouri,  
Columbia; used with permission*

- » Self-expanding or balloon-expandable stents may be used (usually bare metal stents).
- » Complications of percutaneous stenting are in the range of 3% to 7% and include volume overload due to sudden increase in preload, stent thrombosis, pulmonary embolus, stent migration, hematoma at the insertion site, infection, bleeding, and, very rarely, perforation or death.[22]
- » Patency rate is around 80% to 94%, and 20% of patients may require repeat stenting.
- » Bleeding risk is 1% to 14%, due to anticoagulation with aspirin, clopidogrel, and/or warfarin, which may be used following stent placement to prevent thrombosis.[21] [31]
- » Data regarding post-procedural anticoagulation are lacking, and practices vary.[2]

## 2nd

**palliative therapy**

- » Includes palliative radiation therapy, chemotherapy or corticosteroids (for lymphomas and thymomas), endovascular stents, or rarely bypass surgery, as it is invasive and difficult.[1]
- » In rare cases, surgical decompression can be performed.
- » Thrombolysis with indwelling catheters has also been described in small studies.[28]

## Ongoing

» Supportive treatment consists of diuretics, low-salt diet, avoidance of upper-extremity lines, head elevation, and oxygen.

» Type of diuretic will depend on several factors including renal function and current or past history of diuretic use.

## infectious etiology

## 1st treatment of infection

» Underlying infection (e.g., aspergillosis, blastomycosis, histoplasmosis, nocardiosis) should be treated according to local sensitivities.

» Choice of antimicrobial will depend on the underlying pathogen.

## 2nd palliative therapy

» Endovascular stents and more rarely bypass surgery may be required if superior vena cava obstruction persists after treatment of infection.

## iatrogenic etiology

## ■ thrombosis due to central venous catheter(s)

## 1st catheter removal + thrombolysis and/or anticoagulation

» Catheter(s) should be removed and local thrombolysis and/or short-course anticoagulation should be considered in these patients.[29]

» Chronic warfarin therapy up to 1 year has been described in some reports.[32] Dose should be titrated to target INR of 2.0 to 3.0.

» Thrombolysis has higher success rates if used within first 5 days of development of superior vena cava obstruction.[28]

## ■ pacemaker and implantable cardioverter-defibrillator lead-related venous occlusion

## 1st percutaneous balloon dilatation/stenting with or without lead removal

» Percutaneous balloon dilatation/stenting is preferred in these patients.

» Lead explantation may carry a high risk of mortality (1% to 3%).[30] Bypass surgery may be an option.

» Infection of the leads should always be considered as a possibility and evaluated with blood cultures and transesophageal echocardiogram.

» Long-term anticoagulation with warfarin should be considered. Dose should be titrated to target INR of 2.0 to 3.0.

## Primary prevention

The most important primary prevention measure is to avoid smoking, which increases the risk of malignant causes of superior vena cava syndrome.

## Patient discussions

Patients should be advised to monitor for symptoms of recurrence like upper-extremity swelling, engorged neck veins, facial edema, or plethora. They should also be advised to report to the emergency department if they develop dyspnea or confusion.

## Monitoring

### Monitoring

Patients should be followed up regularly by their treating physician (i.e., oncologist/ surgeon/cardiologist/ radiation therapist). The duration, frequency of follow-up, and further workup generally depend on the underlying etiology.

## Complications

Complications	Timeframe	Likelihood
<b>percutaneous stenting procedural complications</b>	<b>short term</b>	<b>low</b>
<p>Procedural complications can include stent thrombosis or migration, dissection, perforation, bleeding, infection, or cardiac tamponade.</p> <p>May require percutaneous or surgical intervention.</p> <p>There is a risk of volume overload or heart failure exacerbation immediately following revascularization due to sudden increase in venous return.</p>		
<b>thrombolysis/anticoagulation-related bleeding</b>	<b>variable</b>	<b>low</b>
<p>Minor or major bleeding complications can be related to systemic anticoagulation. They might necessitate holding the anticoagulant regimen, or possibly giving an antidote.</p>		

## Prognosis

Prognosis usually depends on the underlying etiology, with poorer prognosis for malignant conditions.

### Malignant etiology

In patients with treatment-responsive malignancies, superior vena cava (SVC) syndrome does not necessarily signify adverse outcomes. However, in patients with non-small cell lung cancer resistant to chemotherapy and radiation therapy, development of SVC syndrome is associated with poor prognosis and median survival of <6 months.<sup>[33]</sup>

### Benign etiology

Patients treated for benign causes with stenting or surgery have patency rates of around 90%, though there may be a need for recurrent stenting in some cases. Following percutaneous stenting, patients may need to be on antiplatelet therapy or warfarin for 1 to 3 months, although there are no clear guidelines regarding the duration of treatment.<sup>[22]</sup>



## Diagnostic guidelines

### International

**ACR Appropriateness Criteria: suspected upper-extremity deep vein thrombosis (<https://www.acr.org/Clinical-Resources/Clinical-Tools-and-Reference/Appropriateness-Criteria>) [19]**

**Published by:** American College of Radiology

**Last published:** 2019

## Treatment guidelines

### International

**Antithrombotic therapy for VTE disease: CHEST guideline and expert panel report (<https://www.chestnet.org/Guidelines-and-Resources>) [29]**

**Published by:** American College of Chest Physicians

**Last published:** 2016

## Key articles

- Azizi AH, Shafi I, Shah N, et al. Superior vena cava syndrome. JACC Cardiovasc Interv. 2020 Dec 28;13(24):2896-2910. [Full text \(https://www.sciencedirect.com/science/article/pii/S1936879820319725?via%3Dihub\)](https://www.sciencedirect.com/science/article/pii/S1936879820319725?via%3Dihub) [Abstract \(http://www.ncbi.nlm.nih.gov/pubmed/33357528?tool=bestpractice.bmj.com\)](http://www.ncbi.nlm.nih.gov/pubmed/33357528?tool=bestpractice.bmj.com)
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- Kearon C, Akl EA, Ornelas J, et al. Antithrombotic therapy for VTE disease: CHEST guideline and expert panel report. Chest. 2016 Feb;149(2):315-52. [Full text \(http://journal.chestnet.org/article/S0012-3692\(15\)00335-9/fulltext\)](http://journal.chestnet.org/article/S0012-3692(15)00335-9/fulltext) [Abstract \(http://www.ncbi.nlm.nih.gov/pubmed/26867832?tool=bestpractice.bmj.com\)](http://www.ncbi.nlm.nih.gov/pubmed/26867832?tool=bestpractice.bmj.com)

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## Images

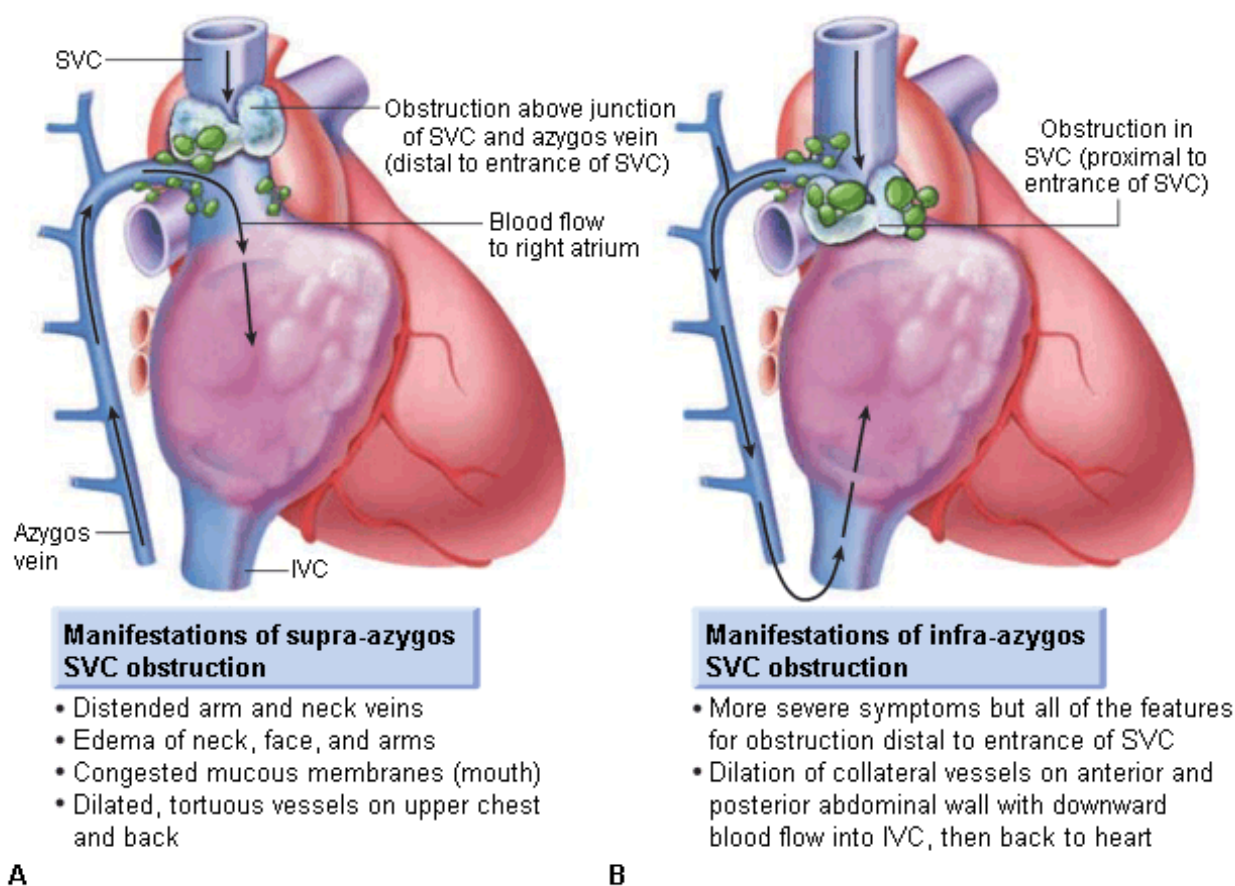
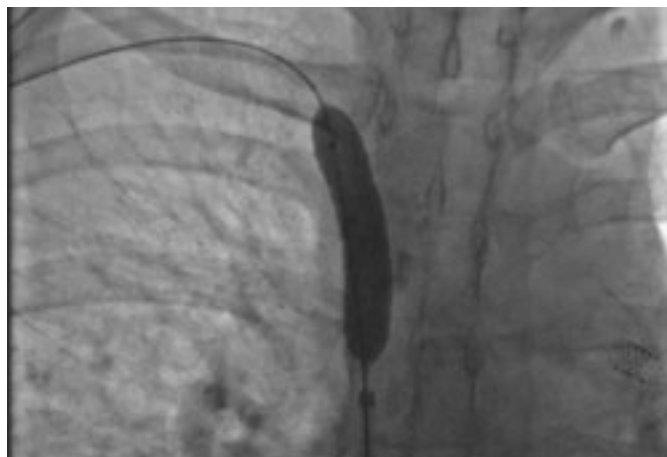


Figure 1: Supra- and infra-azygos obstruction leading to superior vena cava (SVC) syndrome. IVC: inferior vena cava

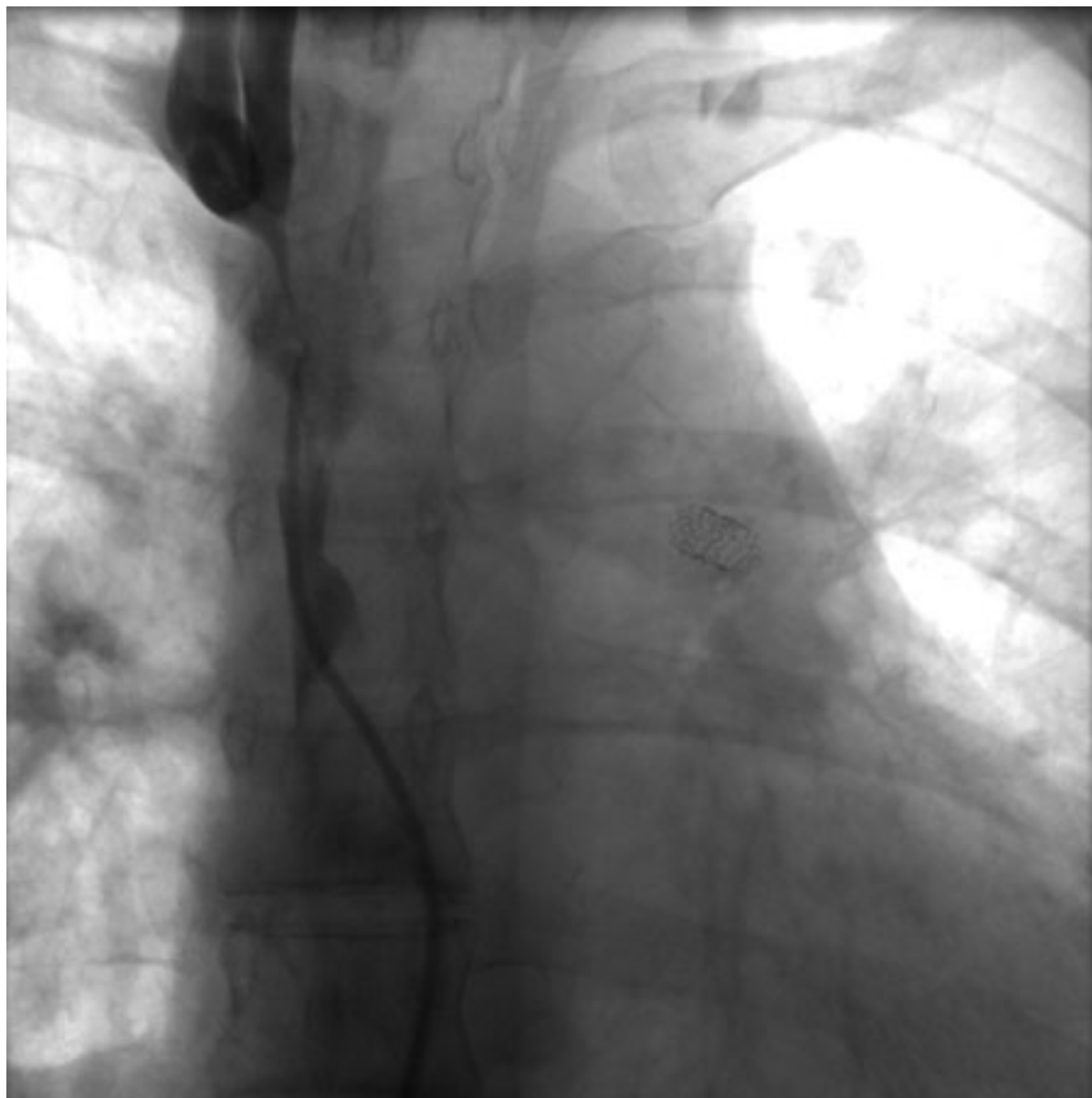
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**Figure 2: Postdilatation of the superior vena cava stent**

*Image obtained from cardiac catheterization laboratory at University of Missouri, Columbia;#used with permission*



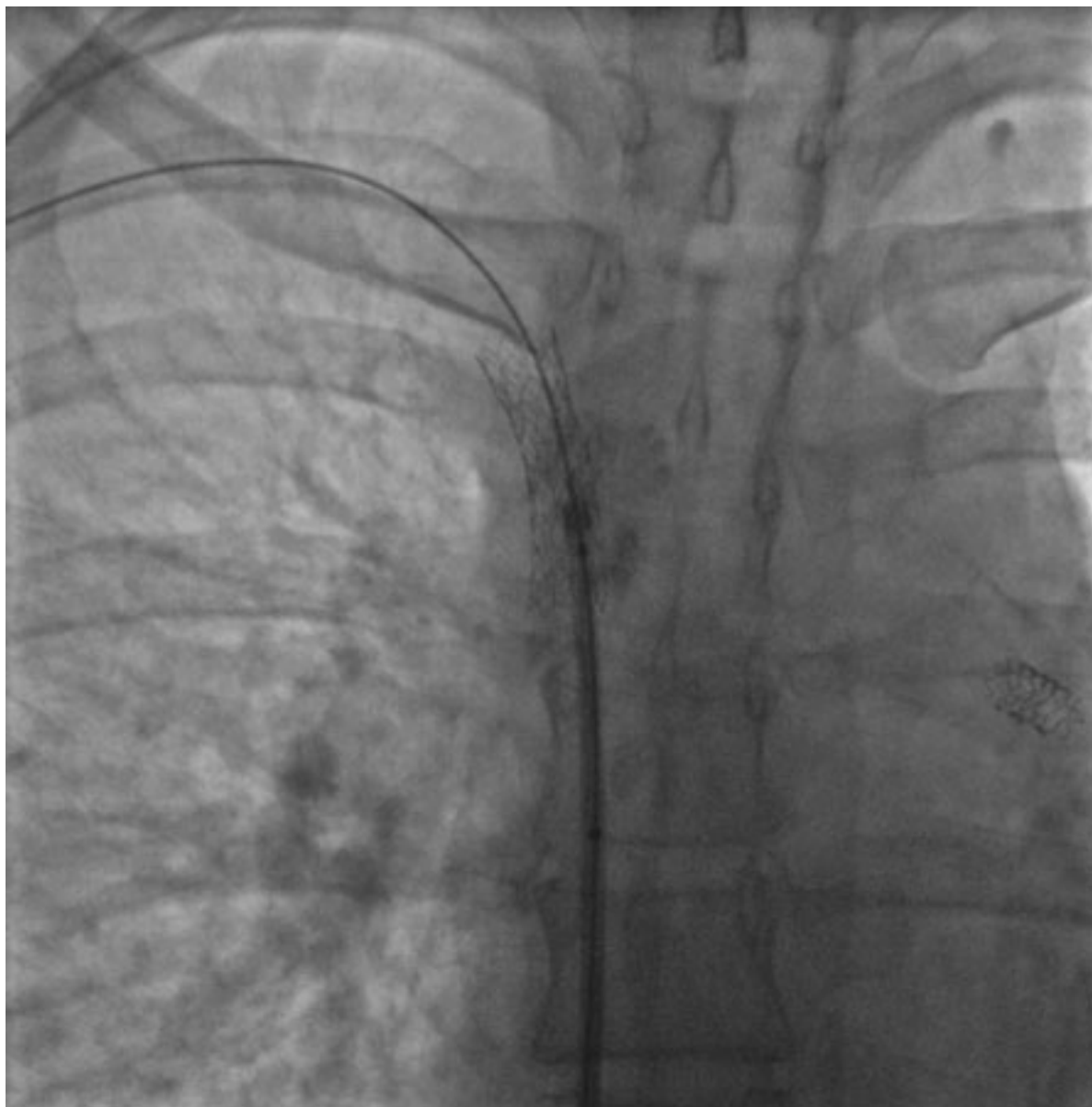
*Figure 3: Venography showing superior vena cava stenosis. Stent placement in the left pulmonary artery is seen*

*Image obtained from cardiac catheterization laboratory at University of Missouri, Columbia; used with permission*



*Figure 4: Percutaneous balloon angioplasty of the stenotic lesion in superior vena cava*

*Image obtained from cardiac catheterization laboratory at University of Missouri, Columbia; used with permission*



**Figure 5: Stent deployment in the superior vena cava**

*Image obtained from cardiac catheterization laboratory at University of Missouri, Columbia; used with permission*

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### Figure 1 – BMJ Best Practice Numeral Style

5-digit numerals: 10,000

4-digit numerals: 1000

numerals < 1: 0.25

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DISCLOSURES: SWY declares that he has no competing interests.

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