BMJ Best Practice

Superior vena cava syndrome

The right clinical information, right where it's needed

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Summary

- Clinical condition that occurs due to obstruction of the superior vena cava.
- Most common aetiology is malignancy; however, there has been an increase in benign causes due to more frequent use of intravascular devices.
- Although rarely fatal, may sometimes present as life-threatening upper airway obstruction.
- High index of suspicion is required to make the diagnosis in many cases.
- Treatment and prognosis depend on underlying aetiology.

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 oedema of the head, neck, and arms, often with cyanosis, plethora, and distended subcutaneous vessels.[1] It can be caused by either intraluminal obstruction of the SVC or extrinsic compression.

Epidemiology

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 resynchronisation therapy.[6] SVC syndrome occurs in approximately 15,000 people in the US every year.[1] Malignancy causes 65% of cases, most commonly lung cancer and non-Hodgkin's lymphoma. Malignant causes of SVC syndrome are more frequent in middle-aged to elderly men, while benign causes are equally distributed across both genders, although more commonly in younger people. Infectious causes (especially syphilitic aortic aneurysm and tuberculosis) accounted for the majority of cases 50 years ago, but are now rare, especially in the developed countries.[7]

Aetiology

A total of 65% of cases are due to malignancy. Lung cancer is the most common aetiology, non-small cell lung cancer accounting for 50% of cases of malignant SVC syndrome and small cell lung cancer for 25% cases.[8] [9] [10] Overall, 2% to 5% of patients with lung cancer go on to develop SVC syndrome; however, 10% to 20% of patients with small cell lung cancer develop SVC syndrome. This is most probably related to the central growth of these tumours.[11] 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, 57% developed SVC syndrome. Although Hodgkin's lymphoma often involves the mediastinum, SVC syndrome is rare.[12] Thymoma (2%) and germ cell tumours (3%) are other primary mediastinal malignancies that occasionally cause SVC syndrome. The most common metastatic disease that causes SVC syndrome is breast cancer, accounting for 11% of cases.[5] Other metastatic tumours that cause SVC syndrome include colon cancer, oesophageal cancer, Kaposi's sarcoma, and fibrous mesothelioma.

Benign causes of SVC syndrome are less frequent (35%). They include iatrogenic causes associated with SVC thrombosis (e.g., central venous catheters, pacemaker and implantable cardioverter-defibrillator leads), mediastinal fibrosis caused by radiotherapy or infections (e.g., histoplasmosis, tuberculosis, aspergillosis, blastomycosis, or nocardiosis), collagen-vascular diseases like sarcoidosis or Behcet's syndrome, and, rarely, aortic arch aneurysm, large substernal goitre, mediastinal haematoma as a result of trauma, and bicaval anastomotic stenosis following cardiac transplantation.[1] [10] [13] 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's 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 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 side of the heart (atrium). It is located in the middle mediastinum and 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 azygous and internal mammary venous system (which takes several weeks).[14] With obstruction of the SVC, the cervical venous pressure is usually increased to 20 to 40 mmHg (normal range 2-8 mmHg).[15] 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 4 principal pathways. The most important pathway is the azygous venous system, which includes the azygous vein, the hemiazygous 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

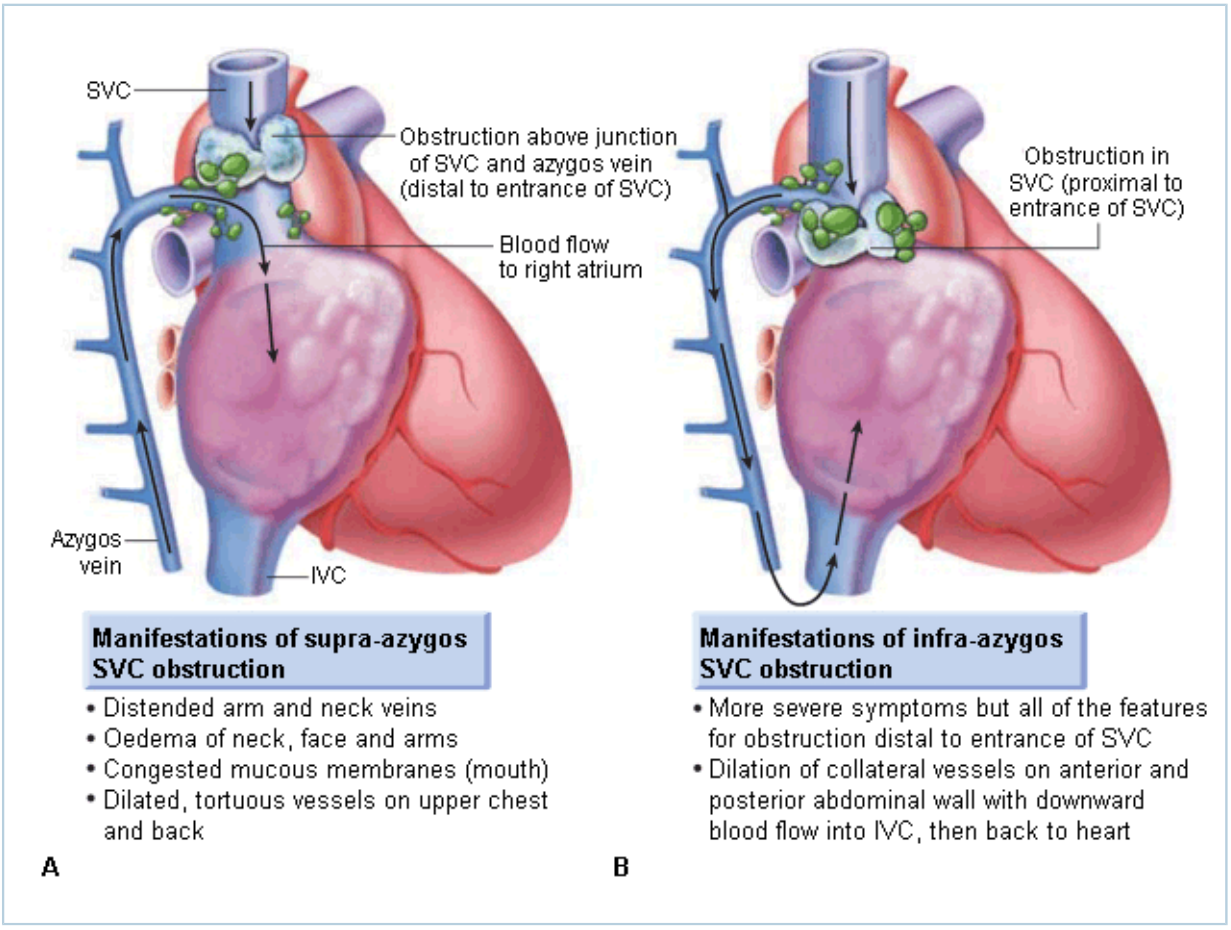
Classification based on location of SVC obstruction

Pre-azygous or supra-azygous

- Obstruction of blood return above the entrance of azygous vein into the SVC, resulting in venous distension and oedema of the face, neck, and upper extremities.[2] [3]

Post-azygous or infra-azygous

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



Supra- and infra-azygous 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 aetiology 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, haematoma, or goitre).

Primary prevention

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

Case history

Case history #1

A 65-year-old man with history of chronic smoking for 40 years, hypertension, and chronic obstructive pulmonary disease presents with anorexia and weight loss for the past 6 months. He had been complaining of increased dyspnoea with exertion and orthopnoea, 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 oedematous, and there were engorged veins in his neck and upper extremity.

Case history #2

A 70-year-old woman, with a history of ischaemic cardiomyopathy, left ventricular ejection fraction of 25%, and prior history of cardiac resynchronisation 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 dyspnoea due to laryngeal oedema that can be acutely fatal due to upper airway compromise.[4] Laryngeal oedema, cyanosis, papilloedema, cerebral oedema, mental changes, stupor, and even coma have been described with severe obstruction.[1] [5] However, these presentations are uncommon in practice and rarely require urgent intervention.

Step-by-step diagnostic approach

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

History

History may also be notable for smoking or multiple pacemaker leads. 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, dyspnoea, cough, arm swelling, and facial plethora.[1] Sudden-onset dyspnoea due to laryngeal oedema can be fatal as a result of upper airway compromise.[4] Other symptoms include headache, chest pain, blurred vision, hoarseness of voice, and stridor. Symptoms are usually worsened by bending forwards or lying down.

Lung malignancy is the most likely aetiology in patients aged >50 years. History of associated anorexia, weight loss, cough, dyspnoea, and haemoptysis 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 oedema, and upper-extremity oedema. Prominent collateral veins covering the anterior chest wall may be visible. The manoeuvre of bending forwards usually worsens venous engorgement and is a helpful clinical sign. Laryngeal oedema, cyanosis, papilloedema, mental changes, stupor, and even coma have been described with severe obstruction.[1] [5] 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 radiological investigation when diagnosis is clinically suspected is chest CT (with intravenous contrast). 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.

Ultrasound of the upper extremities is a useful non-invasive screening test and helps in identification of venous thrombosis or obstruction. 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 in the current era due to improvements in CT and MRI.

Other investigations

Obtaining a tissue diagnosis is important to confirm the presence of malignancy. A 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. 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%.[1]

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 cytological analysis should be strongly considered whenever pleural effusion is present.

ESR or C-reactive protein may be elevated in patients with infection or immunological disorders.

Risk factors

Strong

smoking

- Strong relationship to lung cancer, the most common overall cause of SVC syndrome.

multiple pacemaker leads

- Becoming an increasingly frequent benign cause of SVC syndrome.

Weak

age >50 years

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

radiation

- Excess radiation to the mediastinum can lead to fibrosis causing SVC obstruction.

History & examination factors

Key diagnostic factors

presence of risk factors (common)

- Key risk factors include smoking history and multiple pacemaker leads.

localised oedema of the face and upper extremities (common)

- Present in 80% of cases.[8]
- If oedema is localised to upper extremities and face, obstruction of the SVC should be considered.

dyspnoea (common)

- Present in 60% of cases.[16]
- Usually made worse by bending forwards or lying down (orthopnoea).
- May suggest lung malignancy or chronic infection.

facial plethora (common)

- Due to venous engorgement and oedema.

cough (common)

- Present in 54% of cases.[9]
- Can be related to underlying aetiology or laryngeal oedema.

distended neck veins (common)

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

hoarseness of voice (common)

- Present in 17% of cases.[5]
- Can be related to underlying aetiology or laryngeal oedema.

lymphadenopathy (common)

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

blurred vision (uncommon)

- Present in 2% of cases.[5]

stridor (uncommon)

- Present in 4% of cases.[5]
- Related to laryngeal oedema or direct compression.

confusion/stupor (uncommon)

- Present in 4% of cases and due to cerebral oedema.
- Has been described with severe obstruction.[1] [5]

Other diagnostic factors

anorexia (common)

- May suggest lung malignancy or chronic infection.

weight loss (common)

- May suggest lung malignancy or chronic infection.

haemoptysis (common)

- May suggest lung malignancy or chronic infection.

headache (uncommon)

- Present in 9% of cases.[5]
- 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] [5]

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 oedema (uncommon)

- Has been described with severe obstruction.[1] [5]

cyanosis (uncommon)

- Has been described with severe obstruction.[1] [5]

papilloedema (uncommon)

- Has been described with severe obstruction.[1] [5]

coma (uncommon)

- Has been described with severe obstruction.[1] [5]

Diagnostic tests

1st test to order

Test	Result
chest x-ray • Ordered when SVC syndrome is clinically suspected, especially with a history of pulmonary symptoms.	**widened mediastinum or mass lesion in the lung**
chest CT • Most useful imaging test. • Done with intravenous contrast. • Ordered when there is clinical suspicion of SVC syndrome. • Helps establish diagnosis; shows exact location, severity, and associated pathology (e.g., malignancy or intravascular thrombosis). • Helpful in obtaining a tissue diagnosis by CT-guided biopsy.	**full or partial obstruction; development of collateral vessels; shows location, severity, and pathology**
chest MRI • Useful in patients with a history of contrast allergy or those at risk of contrast-induced worsening of renal function. • Caution advised in use of gadolinium in renal insufficiency due to risk of nephrogenic fibrosing dermopathy. • Contraindicated in patients with pacemakers and defibrillators.	**full or partial obstruction; development of collateral vessels; shows location, severity, and pathology**
ultrasound of upper extremities • Useful non-invasive screening test. • Helps in identification of venous thrombosis or obstruction. • Presence of monophasic flow in the SVC or loss of respiratory variation on Doppler ultrasound can suggest SVC obstruction.	**dilated SVC; presence of thrombus; monophasic flow; loss of respiratory variation**

Other tests to consider

Test	Result
venography • Invasive test, usually performed by venous catheterisation through the femoral vein and injection of contrast dye in the SVC. • Does not provide information about lung or mediastinal pathology.[1] • Not usually required for diagnosis due to improvements in CT and MRI, but useful for endoscopic interventions.	**defines site and extent of SVC obstruction and collateral pathways**

Test	Result
biopsy • Obtaining tissue diagnosis is important to confirm presence 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%.[1] • Biopsy from supraclavicular or other cervical lymph node may obviate the need for invasive procedures such as mediastinoscopy and, thus, careful examination for cervical lymphadenopathy should be performed.	**specimen for pathological diagnosis**
sputum cytology • Simple, non-invasive method to detect lung malignancy. More likely to be positive with central lesions than with peripheral lesions. • Thoracentesis with cytological analysis should be strongly considered when pleural effusion is present.	**malignant cells in sputum**
thoracentesis • Thoracentesis involves placing a needle between the ribs and into the chest to sample fluid that has accumulated in the pleural space. • Thoracentesis with cytological analysis should be strongly considered whenever pleural effusion is present.	**malignant cells in pleural fluid**
sputum culture • Sputum examination for culture is helpful in diagnosis of cases with tuberculosis, or bacterial or fungal infections (e.g., aspergillosis, blastomycosis, histoplasmosis, nocardiosis).	**growth of specific organisms**
ESR • May be present in patients with infection or immunological disorders.	**elevated**
C-reactive protein • May be present in patients with infection or immunological disorders.	**elevated**

Differential diagnosis

Condition	Differentiating signs / symptoms	Differentiating tests
Cardiac tamponade	• Absence of facial and upper-extremity oedema. • Variation of jugular venous pressure (JVP) with respiration (prominent x-descent). • Pulsus paradoxus present.	• Pericardial effusion is seen on CT chest. • Echocardiography shows bouncing septum, marked respiratory variation in the early left ventricular filling velocity (>25%), and right ventricular diastolic collapse.

Condition	Differentiating signs / symptoms	Differentiating tests
Constrictive pericarditis	• Elevated JVP with prominent negative descents (x- and y-descent). • Presence of Kussmaul's sign (increase in JVP with inspiration).	• Echocardiography may show thickened pericardium and marked respiratory variation in the early left ventricular filling velocity (>25%). • MRI is the investigation of choice, as it shows pericardial thickening and ventricular interdependence. • Cardiac catheterisation shows discordance of left and right ventricular pressures with respiration, which has high specificity for diagnosis.
Acute COPD exacerbation	• Extensive bilateral expiratory wheezing, hypoxia, and hypercarbia.	• Peak flow, spirometry, and bronchodilator response help in differentiating. • Presence of obstructive defect on pulmonary function testing is seen.
Right-sided heart failure	• Preserved respiratory variation in JVP, prominent negative descents, and sometimes increased v wave due to tricuspid regurgitation.	• Echocardiography will show right ventricular dysfunction and dilated inferior vena cava with lack of inspiratory collapse.
Pulmonary embolism	• Upper-extremity oedema is usually absent.	• CT chest with contrast will show presence of thrombus inside the pulmonary artery.
Cardiac tumour	• Upper-extremity oedema is usually absent.	• Echocardiography or cardiac MRI will show presence of a mass, usually inside the right side of the heart.

Step-by-step treatment approach

Once diagnosis has been established, a malignant or non-malignant cause of SVC syndrome must be determined, as treatment options differ. Treatment usually involves relieving the symptoms of obstruction and treating the underlying aetiology. There have been no large randomised trials to compare various treatment options, and most data are from case series and expert opinion.

Symptom relief

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 (e.g., acute airway obstruction) if associated with laryngeal or cerebral oedema. This can be achieved with either a combination of corticosteroids and radiotherapy, or percutaneous stenting. Urgent treatment with radiotherapy and corticosteroids should be used only for life-threatening situations. It should be deferred otherwise, due to interference with subsequent histopathological diagnosis. Stenting is 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.[17] [18]

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 tumours causing SVC syndrome are sensitive to radiotherapy. Chemotherapy is an effective option for treatment of lung cancer,[19] lymphomas, and germ cell tumours. Thymomas resistant to chemotherapy and radiation may require surgical resection and SVC reconstruction.[20] Selection of therapy will depend on the type of malignancy, staging, and histopathology.

Second-line treatment is palliative therapy. This includes palliative radiotherapy, chemotherapy or corticosteroids (for lymphomas and thymomas), endovascular stents, or rarely bypass surgery.[1] In rare cases, surgical decompression can be performed. Thrombolysis with indwelling catheters has also been described in small studies.[21] 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 aetiology.

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).

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.[22] Bypass surgery may be an option. Infection of the leads should always be considered as a

possibility and evaluated with blood cultures and transoesophageal echocardiogram. Anticoagulation with warfarin should be considered.

Treatment details overview

Consult your local pharmaceutical database for comprehensive drug information including contraindications, drug interactions, and alternative dosing. (see Disclaimer)

Acute (summary)

Patient group	Tx line	Treatment
acute airway obstruction	1st	secure airway + radiotherapy + corticosteroids
acute airway obstruction	1st	secure airway + percutaneous endovascular stenting

Ongoing (summary)

Patient group	Tx line	Treatment
malignant aetiology	1st	treatment of malignancy
	2nd	palliative therapy
infectious aetiology	1st	treatment of infection
	2nd	palliative therapy
▪ 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 options

Acute

Patient group	Tx line	Treatment
acute airway obstruction	**1st**	**secure airway + radiotherapy + corticosteroids** » Airway should be secured by intubation or surgically first. » Most common cause of SVC obstruction is malignancy, which usually presents with a gradual onset. » Urgent treatment with radiotherapy and corticosteroids should be used only for life-threatening situations. It should be deferred otherwise, due to interference with subsequent histopathological diagnosis. » Dose of corticosteroids should be limited and the dose decreased after 1 to 2 days of treatment or following symptomatic improvement. **Primary options** » radiotherapy **-and-** » dexamethasone: 10 mg intravenous bolus initially, followed by 4 mg every 6 hours
acute airway obstruction	**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.[17] [18] » 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. [Fig-2] [Fig-3] [Fig-4]

Acute

Patient group	Tx line	Treatment
		[Fig-5] » 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, haematoma at the insertion site, infection, bleeding, and, very rarely, perforation or death.[18] » Patency rate is around 80% to 94%, and 20% 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.[17] [23]

Ongoing

Patient group	Tx line	Treatment
malignant aetiology	**1st**	**treatment of malignancy** » Most malignant tumours causing SVC syndrome are sensitive to radiotherapy. » Chemotherapy is an effective option for treatment of lung cancer, lymphomas, and germ cell tumours. » 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%).[20] » Selection of therapy will depend on the type of malignancy, staging, and histopathology.
	2nd	**palliative therapy** » Includes palliative radiotherapy, 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.

Ongoing

Patient group	Tx line	Treatment
		» Thrombolysis with indwelling catheters has also described in small studies.[21] » 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 aetiology	**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 underlying pathogen.
	2nd	**palliative therapy** » Endovascular stents and more rarely bypass surgery may be required if SVC obstruction persists after treatment of infection.
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. » Chronic warfarin therapy up to 1 year has been described in some reports.[24] 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 SVC obstruction.[21]
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%).[22] Bypass surgery may be an option. » Infection of the leads should always be considered as a possibility and evaluated

Ongoing

Patient group	Tx line	Treatment
		with blood cultures and transoesophageal echocardiogram. » Long-term anticoagulation with warfarin should be considered. Dose should be titrated to target INR of 2.0 to 3.0.

Recommendations

Monitoring

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

Patient instructions

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

Complications

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

Prognosis

Prognosis usually depends on the underlying aetiology, with poor prognosis for malignant conditions.

Malignant aetiology

In patients with treatment-responsive malignancies, SVC syndrome does not necessarily signify adverse outcome. However, in patients with non-small cell lung cancer resistant to chemotherapy and radiotherapy, development of SVC syndrome is associated with poor prognosis and median survival of <6 months.[25]

Benign aetiology

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.[18]

Diagnostic guidelines

North America

ACR Appropriateness Criteria: upper extremity swelling

Published by: American College of Radiology **Last published:** 2014

Summary: Guidelines on the use of radiological imaging in patients with suspected upper-extremity deep vein thrombosis.

Treatment guidelines

Europe

Quality assurance guidelines for superior vena cava stenting in malignant disease

Published by: Cardiovascular and Interventional Radiological Society of Europe **Last published:** 2015

Summary: The guideline recommends superior vena cava stenting for patients with malignant obstruction.

Guidance on use of vena cava filters

Published by: British Committee for Standards on Haematology **Last published:** 2006

Summary: The guideline makes recommendations on appropriate use of vena cava filters.

Stent placement for vena caval obstruction

Published by: National Institute for Health and Care Excellence **Last published:** 2004

Summary: NICE supports the use of stent placement for vena caval obstruction.

North America

Antithrombotic therapy for VTE disease: CHEST guideline and expert panel report

Published by: American College of Chest Physicians **Last published:** 2016

Summary: Evidence-based guidelines on the use of thrombolysis and anticoagulation in patients with venous thromboembolism.

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Images

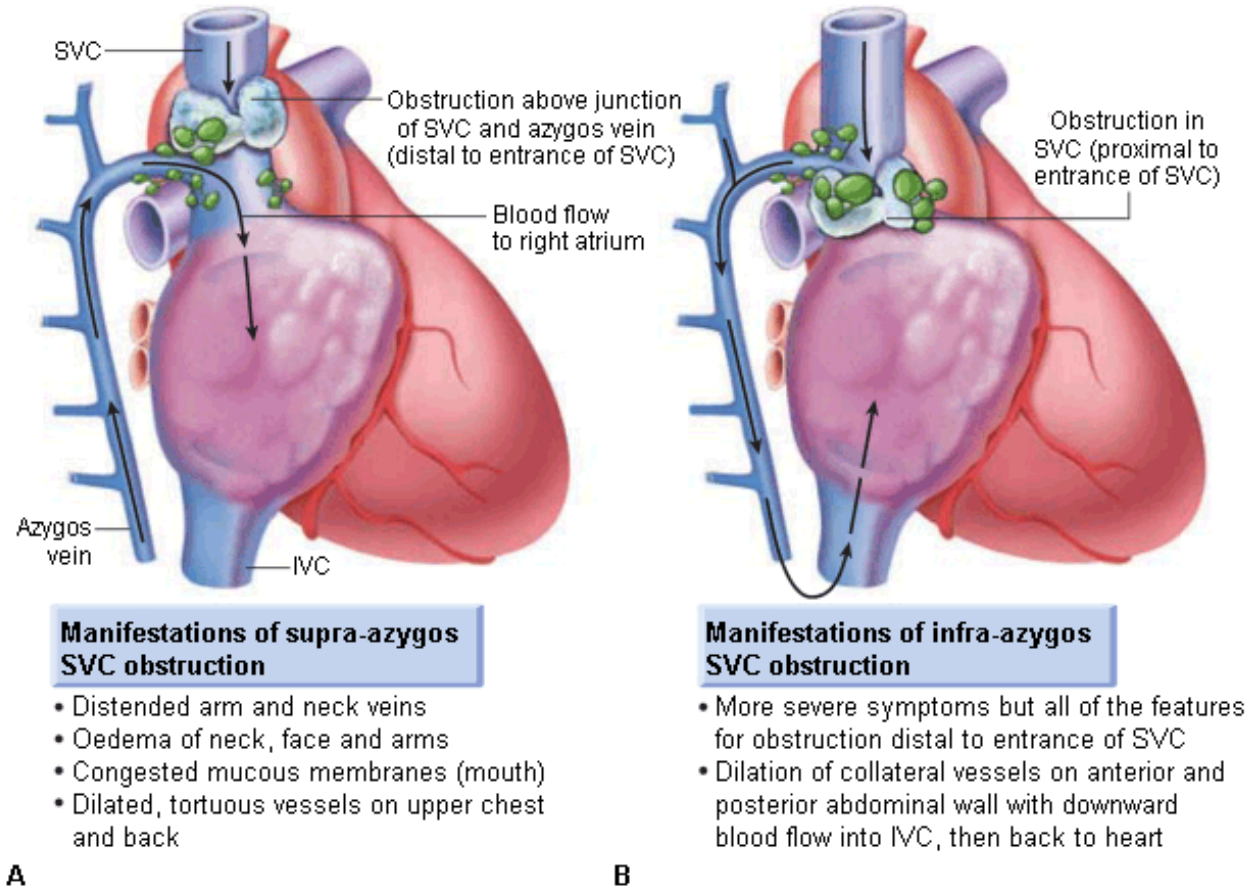

Figure 1: Supra- and infra-azygous obstruction leading to superior vena cava (SVC) syndrome. IVC: inferior vena cava

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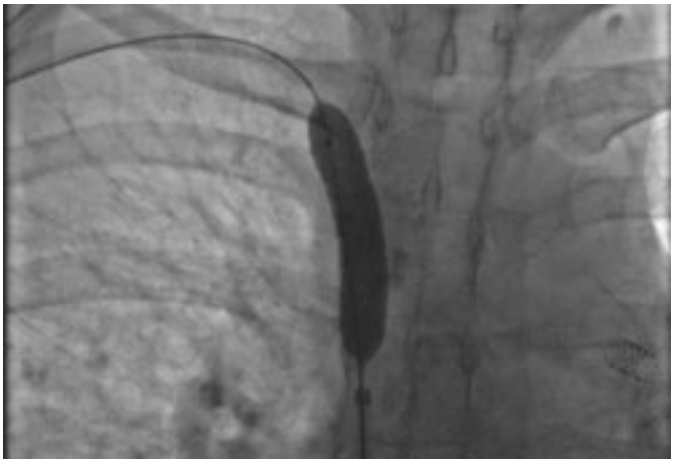

Figure 2: Post-dilatation of the superior vena cava stent

Image obtained from cardiac catheterisation laboratory at University of Missouri, Columbia

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

Image obtained from cardiac catheterisation laboratory at University of Missouri, Columbia

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

Image obtained from cardiac catheterisation laboratory at University of Missouri, Columbia

Figure 5: Stent deployment in the superior vena cava

Image obtained from cardiac catheterisation laboratory at University of Missouri, Columbia

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