

SUPERIOR VENA CAVA SYNDROME

WHAT IS SUPERIOR VENA CAVA SYNDROME?

Superior vena cava syndrome (SVCS) is a collection of clinical signs and symptoms resulting from either partial or complete obstruction of blood flow through the superior vena cava (SVC). The obstruction is most commonly a result of tumor infiltration or thrombus formation in the vessel wall.¹

The SVC is formed by the junction of the left and right innominate (brachiocephalic) veins and is tasked with returning blood from the head, neck, upper extremities, and torso back to the heart. Venous congestion can produce a clinical scenario of increased upper body venous pressures.¹

SIGNS & SYMPTOMS

Intrathoracic malignancy causes 60%–85% of cases of SVCS, usually non-small cell lung cancer (50% of the time), small cell lung cancer (25%–35%), and non-Hodgkin lymphoma (10%–15%). Other causes include thrombus and stenosis secondary to venous lines.² Research shows, venous access devices account for 15%–40% of SVC obstructions in non-malignant conditions.³

Symptoms of SVCS include swelling of the nose, face, neck, or upper extremities; dyspnea; cough; lethargy; a feeling of fullness in the head; abnormally rapid breathing while the patient is lying down or bending forward; and dilated chest vein collaterals.^{1,2}

Symptoms may be most apparent in the morning, after the patient has been in a recumbent position for some time.² When the collateral veins cannot accommodate the excess pressure, the frequency of the symptoms increases.² Symptoms may vary depending upon the degree of the obstruction, development of collateral circulation, and underlying etiology.

Recognizing early signs of SVCS allows for accurate diagnosis, as well as identification of treatment options and goals of care. Discussions about goals of care should occur prior to advanced symptoms, which include respiratory and neurological distress.

Hemodynamic Symptoms³	Respiratory Symptoms³	Neurological Symptoms³
Facial or neck edema Upper-extremity edema Chest pain Distended neck veins Distended chest veins Facial plethora Visual symptoms Cyanosis	Dyspnea (most common presenting symptom) Cough Hoarseness Stridor or central airway obstruction (medical emergency) Severe laryngeal edema (medical emergency)	Headache Dizziness Confusion Obtundation or cerebrovascular accident (medical emergency) Coma from cerebral edema (medical emergency)

INTERVENTIONS

- Manage medical emergencies associated with SVCS in accordance with the patient's and family's goals of care (e.g., secure airway, support breathing and circulation).
- Recognize early signs of SVCS to allow treatment before life-threatening symptoms of respiratory and neurologic distress occur.
- Focus on identifying the underlying etiology of SVCS

Increased predominance of SVCS caused by chronic diseases has prompted the development of new treatment methods. Stenting of the SVC has become widely accepted as a palliative treatment for SVCS associated with malignant disease (endovenous recanalization with stent placement).³

NONPHARMACOLOGICAL INTERVENTIONS²	PHARMACOLOGICAL INTERVENTIONS²
<p><u>Reduce symptom burden:</u></p> <p>Elevate patient's head, as tolerated, to decrease hydrostatic pressure and head and neck edema.</p> <p>Administer oxygen therapy where available.</p> <p>Teach energy conservation.</p>	<p><u>Thrombolytic therapy:</u></p> <p>Tissue plasminogen medication is the most commonly used thrombolytic medication. It should be initiated within two to five days of initial symptoms for optimal outcomes and may not be effective after the 10th day. Costs may be prohibitive in hospice settings.</p>

Closely monitor fluid and electrolyte balance, as overhydration may exacerbate the symptoms of SVCS.

Avoid blood pressure measurements in the upper extremities.

Diagnostics:

Diagnostics may be used to identify the extent of obstruction and damage, as well as assist in the development of a treatment plan. Cost may be prohibitive in hospice settings.

- **Chest x-ray** is the most common diagnostic study performed at initial presentation to evaluate for mass, pleural effusion, or widening of the mediastinum.
- **Venography** is the gold standard for visualizing and diagnosing a venous obstruction. It is used during potential endovascular treatments (such as stenting) depending on the extent and urgency of the illness.
- **Ultrasound** of the jugular, subclavian, and innominate veins can help identify a thrombus within the vessel lumen.
- **Radiographic imaging and magnetic resonance imaging** may be needed to identify the location, severity, and etiology of the SVC obstruction.
- **Chest computed tomography (CT)**, when collateral vessels are present, has a high sensitivity to provide diagnostic clarity. CT scan can clarify the extent of the cavity damage and whether thoracic surgery may be necessary.

Drug therapy:

- **Chemotherapy or radiation therapy** may be used when a tumor is the cause of the obstruction. Cost may be prohibitive in hospice settings.
- **Anti-inflammatory steroids** such as dexamethasone and diuretics have been used to treat SVCS despite a shortage of research to support their effectiveness.
- **Avoid overhydration.**

FAMILY & TEAM DISCUSSIONS

Patient and Family Education and Support²:

- Assess the patient's unique coping needs and provide individualized support.
- Educate the patient and family about appropriate nonpharmacological and pharmacological strategies, including elevated head positioning and energy conservation.
- Provide education on underlying etiology of SVCS, treatment options, benefits and burdens of invasive versus noninvasive management, medication adverse reactions, and indicators of response to therapy for reporting purposes.
- Clarify the patient's and family's goals and wishes, as well as their definition of quality of life, when evaluating treatment options.

Interprofessional Team:

Successful interventions in caring for patients with SVCS benefit from multiple perspectives to anticipate, prevent, and treat physical, psychological, social, and spiritual needs.

Consider social work, psychology, counseling, and spiritual care specialists for palliative and hospice support, as well as interventions to address concerns regarding caregiver support, fear, anxiety, guilt, depression, spiritual and cultural rituals, and financial concerns, as culturally appropriate.

SYMPTOM DOCUMENTATION EXAMPLE

59 yr old female with stage 4 non-small cell lung cancer (NSCLC), current PPS (Palliative Performance Scale) of 40% with progressive functional decline over the 3 months. Over the past week, the patient experienced worsening shortness of breath and increased swelling in her face, which resulted in the patient going to the emergency room and subsequently being diagnosed with SVCS. Ongoing goals-of-care discussions related to NSCLC disease progression, and SVCS emergency interventions and management discussed with patient and surrogate decision-maker. Patient and family agreed to hospice referral to evaluate options. Hospice referral made. Family meeting scheduled for this afternoon. Plan of care updated to reflect hospice evaluation.

DESIRED NURSING OUTCOMES

- Promote informed, shared decision-making regarding treatment interventions for SVCS.
- Assess patient for side effects of SVCS treatment for prompt recognition and intervention.
- Improve physical, psychological, social, and spiritual well-being of patient and caregivers suffering from the distressing symptoms of SVCS and provide interventions.
- Promote ongoing conversations about goals of care relating to disease progression and emergency interventions and management.

REFERENCES

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3. UpToDate. Malignancy-related superior vena cava syndrome. Available at: <https://www.uptodate.com/contents/malignancy-related-superior-vena-cava-syndrome>. Accessed 9/29/20.