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Regional Palliative Care Services

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Contacts:

Northern Health Regional Palliative Care Nurse Consultation Line 250-645-3791

Email: Palliative.Care.Consult.Team@northernhealth.ca

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Superior Vena Cava Syndrome

By Kelsey Lindeman, RN, Palliative Care Nurse Consultant

Superior Vena Cava Syndrome (SVCS) is a distressing and potentially life threatening complication that may arise in palliative care. Early identification and management can successfully reduce mortality and greatly improve outcomes, but because it is a relatively uncommon occurrence, not all clinicians are familiar with identifying and treating SVCS. It is a very treatable condition if identified early, therefore it is important that we are familiar with how to assess and identify patients at risk for SVCS so it can be managed appropriately.

This article will help the primary care clinicians who are involved in the care of palliative patients to be able to identify the signs and symptoms of SVCS, appropriate diagnostic procedures, as well as the treatment options available for symptom management based on underlying etiology and the patient's goals of care.

What is SVC Syndrome?

The superior vena cava (SVC) is a large vein with a thin wall and a low intravascular pressure, surrounded by a constricted compartment made up of the rigid structures of the thorax (Lepper et al., 2011). This makes it difficult for the blood vessel to expand, but easily susceptible to compression. The SVC carries close to one third of the body's total venous return, transporting blood from the head, neck, upper extremities and chest, to the heart. SVCS happens when the blood flow from these areas becomes restricted, causing an increase in pressure and fluid build up in the veins and surrounding areas that drain into the superior vena cava. This restriction of blood flow can be thrombotic (caused by the formation of a blot clot) or caused by tumors in the mediastinum that compress the SVC (Wilson et al., 2007). In this way, SVC obstruction can be caused by either intrinsic or extrinsic obstruction.

The restriction of blood flow through the SVC results in

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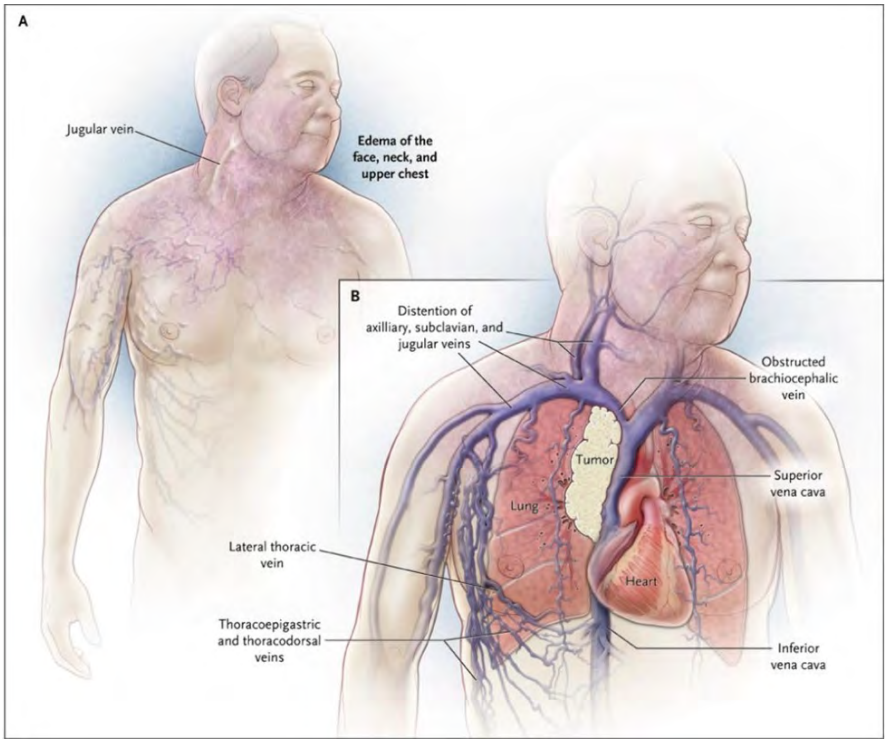
impaired venous return to the heart and usually a gradual onset of classic signs and symptoms. Most patients will display some of the symptoms from 2- 4 weeks before diagnosis. In some cases however, onset is rapid and can threaten airway patency.



Signs and Symptoms

Many early signs can parallel expected symptoms of bronchogenic cancers, such as dyspnea, non-productive cough and hoarseness. In addition to these symptoms, other hallmark signs include dysphagia, sensation of fullness in the face and upper extremities, periorbital edema, neck vein and chest wall vein distention as well as edema of the face and upper extremities. Dilation of veins on the hand may be present and do not collapse when the arm is elevated. The presence of enlarged veins in the upper body is a result of collateral circulation that develops in order to bypass the site of obstruction. This allows the collateral circulation to maintain adequate blood flow to tissues that otherwise would be jeopardized by ischemia related to the reduced blood flow caused by SVC obstruction.

Late signs can include cerebral edema causing headache, irritability, visual disturbances, and changes in level of consciousness. Horner's syndrome (characterized by a constricted pupil (miosis), drooping of the upper eyelid (ptosis), absence of sweating of the face (anhidrosis), sinking of the eyeball into the bony cavity that protects the eye (enophthalmos) and cyanosis of the face or upper torso are rare but may also occur. Tachypnea, and orthopnea may also be present.

Edema in the upper body can be visually striking, but often is not life threatening. Cerebral edema however, although rare, can be serious or fatal. Cerebral edema can lead to brain stem herniation and possibly death. Narrowing of the upper respiratory tract as a result of nasal and laryngeal edema can also become life threatening.

Not all signs described will necessarily be present, and may vary in intensity. A classification system for grading the severity of malignant superior vena cava syndrome was developed to guide SVCS management.

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Grading the severity of malignant superior vena cava syndrome

Grade	Findings	Estimated incidence (%)
0	Asymptomatic – Radiographic superior vena cava obstruction in the absence of symptoms	10
1	Mild – Edema in head or neck (vascular distention), cyanosis, plethora	25
2	Moderate – Edema in head or neck with functional impairment (mild dysphagia, cough, mild or moderate impairment of head, jaw, or eyelid movements, visual disturbances caused by ocular edema)	50
3	Severe – Mild or moderate cerebral edema (headache, dizziness), mild/moderate laryngeal edema, or diminished cardiac reserve (syncope after bending)	10
4	Life-threatening – Significant cerebral edema (confusion, obtundation), significant laryngeal edema (stridor), or significant hemodynamic compromise (syncope without precipitating factors, hypotension, renal insufficiency)	5
5	Fatal – Death	<1

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Graphic 117725 Version 1.0

Incidence and Risk Factors

SVCS occurs in 3-4% of cancer patients. 90% of the cases that do occur are in patients with bronchogenic cancers (non-small cell, small cell and right sided lung cancer) and non-Hodgkin's lymphoma (diffuse large cell, lymphoblastic lymphoma). SVCS tends to be less common, but still occurs, in Hodgkin's lymphoma (with or without mediastinal involvement), metastatic breast cancer, germ cell tumors, thymoma and mesothelioma. Bronchogenic cancers are among the highest risk for developing SVCS (BC Cancer, 2021).

In recent years there has been an increase in the number of SVCS cases seen due to thrombotic complications, which is believed to be a direct reflection of the increased use of central venous access devices and other intravascular devices such as port-a-caths, PICC lines and pacemakers (Ghanavati et al., 2017). This emphasizes that not only cancer patients are at risk and susceptible to SVCS and its complications.

Diagnostics

It is important with any diagnostics to consider the patient's goals of care. Diagnostics are warranted only when an intervention (such as placement of a stent or surgery) is planned and in line with the patient's wishes for treatment. If none of the treatment options listed below are available or acceptable to the patient, it would not be necessary to undergo the diagnostics to determine the most appropriate intervention.

Imaging may be warranted in the presence of some or all of the symptoms listed above, to confirm the presence of SVCS and determine etiology. Treatment decisions will differ depending on etiology.

CT of the chest with contrast is one of the most useful imaging studies, providing information on mediastinal structures and extent of disease and involvement. It also useful in determining the extent of vascular thrombi, pathways of venous circulation and vascular compression or obstruction. It can also be effective at localizing the tumor for CT guided biopsy and/or determining the radiation treatment field.

X-ray can also be useful for indicating presence of a mass, mediastinal widening or pleural effusion, but will not confirm vascular thrombosis. It can be the most accessible imaging study to obtain, however results may be inconclusive if the cause is thrombotic in nature.

Contrast venography and MRI can be useful for visualizing mediastinal structures, circulation pathways and patency.

Treatment

Treatment will depend on diagnostic results, goals of care, functional status, prognosis and whether the underlying cause of SVCS is due to thrombosis or physical compression/obstruction from tumor burden.

Thrombotic therapy is a treatment used to dissolve blood clots in blood vessels, which helps to improve blood flow. In the presence of a

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central venous access device, thrombolytic infusions will be infused directly through the device, however in the absence of such a device thrombolytics can be administered through peripheral venous access devices as well. Options for thrombotic therapy include streptokinase, urokinase and recombinant tissue plasminogen activator (rt-PA).

For SVCS caused by physical compression from tumor burden, radiotherapy, chemotherapy or a combination of both of these treatments may be warranted. Other options include intravascular stent placement, surgical intervention and medical management.

Radiotherapy is most appropriate for patients with known radiosensitive tumors such as lymphomas or non small cell lung cancer. Radiotherapy is delivered by a machine that destroys cells within the treatment area. Cells are damaged directly by DNA destruction, preventing the cells from continuing to grow. Cancer cells are known to be more sensitive to radiation than normal cells, although it is difficult to damage cancer cells without damaging some normal cells as well. Radiation as a treatment option for SVCS and can also be used without a histological diagnosis for those experiencing rapid deterioration due to increased intracranial pressure, or those at risk for airway compromise due to severe airway compression. Treatment burdens to consider that are associated with radiotherapy can include skin reaction, fatigue, esophagitis, cough and nausea. Delayed side effects may also include pneumonitis, pulmonary fibrosis, stenosis or ulceration of the esophagus, cardiac changes, spinal cord myelopathy or brachial plexopathy (damage to the brachial plexus causing peripheral neuropathy). Radiotherapy side effects will depend on the total dose of radiotherapy prescribed, the dose per fraction, the volume of normal tissue in the radiation field, and whether or not chemotherapy is received concurrently. Symptom relief with radiotherapy is possible within 5-10 days of treatment, but may take up to 4 weeks to manifest fully. Relief may last for several months (BC Cancer, 2021).

Chemotherapy may be considered for

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Palliative Care Education in Northern Health

Did you know we have Nurse Consultants for each HSDA?

In September 2021, our Regional Palliative Care Consultation Team travelled to both the Northwest and Northeast to provide palliative care education and network with primary care providers.

Palliative Care Nurse Consultant, Jennifer Krzyczkowski, presenting in the Northwest, September 2021.

malignancies that are highly chemosensitive such as small cell lung cancer and non-Hodgkins lymphoma. Chemotherapy is a large group of systemic drugs that when administered, travel throughout the body by blood stream. Some cancers may only need one kind of chemotherapy, others may need a combination. These drugs are used to destroy cancer cells, but they can affect both cancer and normal cells. SVCS symptom relief with chemotherapy may be between 7-14 days (BC Cancer, 2021). Side effects will depend on the specific chemotherapy agents used, but may include myelosuppression, nausea, vomiting, stomatitis, esophagitis and alopecia. Another complication to consider with administering chemotherapy agents in the context of SVCS is that SVCS causes venous stasis. This can result in an increased local concentration of IV drugs in the upper thorax and can cause poor drug absorption and vessel wall irritation.

Combination therapy may benefit some patients. An initial treatment of chemotherapy is used to help decrease tumor size, followed by radiation with a smaller radiation treatment field. This approach can be ideal if the patient has a high performance status and there is a chance of down-staging a malignancy to a resectable lesion. This combination approach however does result in higher rates of esophagitis and neutropenia.

Intravascular stent placement should be considered for malignancies not particularly sensitive to chemotherapy or radiation. This is a procedure that is performed by an interventional radiologist under local anaesthetic. A stent is inserted via the basilica, subclavian or femoral vein. The stent expands inside the SVC to relieve obstruction, increase the diameter of the vessel and thereby improve the ability for blood to flow through. This treatment requires ongoing

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Updated Contact Information:

To contact our team please dial 250-645-3791. To skip the message press 1 for general questions or 2 for the nurse consultant menu. In the nurse consultant menu, please select a number as per the table below to reach the nurse consultant responsible for your area.
If you are unsure who to contact: please dial 250-645-3791 and press 1 to leave a message.
E-mail: Palliative.Care.Consult.Team@northernhealth.ca or Fax Requests to: 250-565-5596

Nurse Consultant	Areas Covered				
Seth Gysbers Press 1	UHNBC: FMU, PCMU & Emergency	Gateway Assisted Living			
Jennifer Ferguson Press 2	UHNBC: Surgery North & South	McBride	Valemount	Prince George Community Nursing	
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anticoagulation therapy for prophylactic thrombus prevention, but is known to provide relief in up to 90% of patients (Pallium Canada, 2018). This procedure also has very good long-term patency rates, allowing good long term symptom relief. Initial symptom relief also tends to be faster than radiotherapy or chemotherapy, with decreased symptom burden being evident within 24-72 hours of the procedure (Watkinson et al., 2008). Complications of intravascular stent placement include thrombosis formation, stent migration, hematoma development, infection, deep vein thrombosis or pulmonary embolism.

Surgery is less common and largely replaced by intravascular stents due to limited success and high morbidity. Surgical bypass of the SVC is usually less effective in SVCS caused by malignancy than treating the underlying tumor burden and preventing progression or recurrence. Surgery is generally only indicated in the presence of severe symptoms and when less invasive interventions have failed to provide symptom relief.

Medical management can be considered for patients in the home or acute care setting. Positioning the patient for comfort by elevating the head of the bed allows for optimizing oxygenation and patient comfort. Elevating arms may help increase venous return. Oxygen therapy may be effective for relieving dyspnea in hypoxic patients. Remove rings and restrictive clothing for comfort and avoid venipuncture to affected extremities. Stool softeners and cough suppressors may be indicated to help prevent the use of the Valsalva maneuver which increases venous pressure. Medications such as analgesics and anxiolytics may ultimately be necessary to manage pain or anxiety.

Diuretics have been recommended by some guidelines for management of edema and fluid retention (Lepper et al., 2011; BC Cancer, 2021), however effectiveness has also been questioned especially considering the action of diuretics is to decrease the preload which could actually negatively affect cardiac output (Wilson et.al, 2007; Pallium Canada, 2018).

Dexamethasone is a non-invasive intervention that can be considered as either a sole agent in treatment of SVCS, or as a temporary measure that can be used while awaiting diagnostics and/ or more definitive treatment. For some, depending on goals of care, availability of diagnostic and treatment options as well as disease trajectory, dexamethasone may be the most appropriate intervention. Sometimes a trial of dexamethasone alone as initial management, can be sufficient for symptom management of SVCS.

Final Thoughts

Confidently identifying the early and late signs of SVCS is an important clinical skill especially for those working with patient populations at risk for developing this syndrome. Early intervention and management can reduce symptom burden and improve quality of life for patients during the palliative phase.

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Upcoming Palliative Care Education Sessions

The Northern Health Palliative Care Consultation Team is excited to offer a variety of upcoming palliative care education in both in person and online formats. If you are interested or have questions about our upcoming training please e-mail Palliative.Care.Consult.Team@northernhealth.ca.

In Person

Session:	Date:	Time:	Location:	Who can register:
LEAP—Learning Essential Approaches to Palliative Care	April 27 & 28, 2022	08:00 to 16:30	Prince George	RNs & LPNs

Online

Session:	Date:	Time:	Who can register:
ECHO—Basic	March 8, April 5, May 3, June 7	12:00 to 13:00	RNs and LPNs
Serious Illness Conversation Training Guide	April 8, 2022	14:30 to 16:00	Primary Care Providers
Integrating a Palliative Approach: Essentials for Personal Support Workers	Starting April 2022	Online	Personal Support Workers, Community Health Workers, and Care Aides
Essentials in Hospice and Palliative Care: A Practical Resource for Every Nurse	Starting April 2022	Online	RNs and LPNs