

**American College of Radiology
ACR Appropriateness Criteria®**

Thoracic Venous Occlusions-Suspected Superior Vena Cava Syndrome

Variant: 1 Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

| Procedure | Appropriateness Category | Relative Radiation Level |
|--|-----------------------------------|--------------------------|
| MRA and MRV chest without and with IV contrast | Usually Appropriate | O |
| MRV chest without and with IV contrast | Usually Appropriate | O |
| CT chest with IV contrast | Usually Appropriate | ⊕⊕⊕ |
| CT chest without and with IV contrast | Usually Appropriate | ⊕⊕⊕ |
| CT neck and chest with IV contrast | Usually Appropriate | ⊕⊕⊕⊕ |
| CT neck and chest without and with IV contrast | Usually Appropriate | ⊕⊕⊕⊕ |
| CTA and CTV chest with IV contrast | Usually Appropriate | ⊕⊕⊕⊕ |
| CTV chest with IV contrast | Usually Appropriate | ⊕⊕⊕⊕ |
| Radiography chest | May Be Appropriate (Disagreement) | ⊕ |
| MRA chest with IV contrast | May Be Appropriate (Disagreement) | O |
| MRA chest without and with IV contrast | May Be Appropriate (Disagreement) | O |
| MRA chest without IV contrast | May Be Appropriate | O |
| MRI chest with IV contrast | May Be Appropriate | O |
| MRI chest without and with IV contrast | May Be Appropriate | O |
| MRI chest without IV contrast | May Be Appropriate | O |
| MRV chest without IV contrast | May Be Appropriate (Disagreement) | O |
| CTA chest with IV contrast | May Be Appropriate (Disagreement) | ⊕⊕⊕ |
| CTA chest without and with IV contrast | May Be Appropriate (Disagreement) | ⊕⊕⊕ |
| US duplex Doppler upper extremity and chest | Usually Not Appropriate | O |
| Catheter venography upper extremity and SVC | Usually Not Appropriate | ⊕⊕⊕ |
| CT chest without IV contrast | Usually Not Appropriate | ⊕⊕⊕ |
| CT neck and chest without IV contrast | Usually Not Appropriate | ⊕⊕⊕⊕ |

Variant: 2 Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

| Procedure | Appropriateness Category | Relative Radiation Level |
|--|-----------------------------------|--------------------------|
| MRA and MRV chest without and with IV contrast | Usually Appropriate | O |
| MRV chest without and with IV contrast | Usually Appropriate | O |
| CT chest with IV contrast | Usually Appropriate | ⊕⊕⊕ |
| CT chest without and with IV contrast | Usually Appropriate | ⊕⊕⊕ |
| CT neck and chest with IV contrast | Usually Appropriate | ⊕⊕⊕⊕ |
| CT neck and chest without and with IV contrast | Usually Appropriate | ⊕⊕⊕⊕ |
| CTA and CTV chest with IV contrast | Usually Appropriate | ⊕⊕⊕⊕ |
| CTV chest with IV contrast | Usually Appropriate | ⊕⊕⊕⊕ |
| US duplex Doppler upper extremity and chest | May Be Appropriate (Disagreement) | O |
| Radiography chest | May Be Appropriate (Disagreement) | ⊕ |

| | | |
|---|-----------------------------------|------|
| Catheter venography upper extremity and SVC | May Be Appropriate (Disagreement) | ⊕⊕⊕ |
| MRA chest with IV contrast | May Be Appropriate | ○ |
| MRA chest without and with IV contrast | May Be Appropriate | ○ |
| MRI chest with IV contrast | May Be Appropriate | ○ |
| MRI chest without and with IV contrast | May Be Appropriate | ○ |
| MRI chest without IV contrast | May Be Appropriate | ○ |
| MRV chest without IV contrast | May Be Appropriate (Disagreement) | ○ |
| CTA chest with IV contrast | May Be Appropriate (Disagreement) | ⊕⊕⊕ |
| CTA chest without and with IV contrast | May Be Appropriate (Disagreement) | ⊕⊕⊕ |
| MRA chest without IV contrast | Usually Not Appropriate | ○ |
| CT chest without IV contrast | Usually Not Appropriate | ⊕⊕⊕ |
| CT neck and chest without IV contrast | Usually Not Appropriate | ⊕⊕⊕⊕ |

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Summary of Literature Review

Introduction/Background

Venous drainage of the head, neck, and upper extremities returns to the heart via the superior vena cava (SVC). Obstruction of the SVC can occur from a variety of causes, including intrinsic obstructive and extrinsic compressive pathologies. A review of the causes, diagnosis, and management of SVC syndrome is presented here in addition to an evaluation of relevant imaging studies.

SVC syndrome occurs in approximately 15,000 people in the United States each year. It most commonly occurs secondary to thoracic malignancies, mostly primary lung cancer and lymphoma. Even with the increased use of intravascular devices, the prevalence of SVC thrombosis or stenosis related to these devices is increasing [1]. Other benign causes include fibrosing mediastinitis, tuberculosis, histoplasmosis, thyroid goiter, and aortic aneurysms [1,2]. Symptoms typically present over weeks to months, although they can also present acutely over minutes to hours. The incidence of acute SVC syndrome is not well documented, although it is considered relatively rare compared with chronic or subacute forms [2,3].

SVC syndrome is typically not acutely life-threatening, and management revolves around relieving symptoms and treating the underlying etiology. Treatments can include elevating the patient's head, anticoagulation, diuretics, chemotherapy, thrombolysis, thrombectomy, endovascular recanalization with stent placement, and/or radiation therapy [2-4].

Initial Imaging Definition

Initial imaging is defined as imaging at the beginning of the care episode for the medical condition defined by the variant. More than one procedure can be considered usually appropriate in the

initial imaging evaluation when:

- There are procedures that are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care)

OR

- There are complementary procedures (ie, more than one procedure is ordered as a set or simultaneously wherein each procedure provides unique clinical information to effectively manage the patient's care).

Discussion of Procedures by Variant

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

SVC syndrome arises from compromised venous return from the head, neck, and upper extremities, typically evolving gradually as collateral circulation develops over time. However, in cases of acute thrombosis or embolism, symptoms may abruptly manifest before collateral circulation is established. Common symptoms include facial and neck swelling, upper extremity discoloration, dyspnea, cough, dysphagia, dizziness, blurred vision, breathing difficulty, and hoarseness. Although symptoms often cause significant discomfort, they are typically not life-threatening. Nevertheless, in severe cases of increased venous pressures, altered mental status, hypotension, life-threatening cerebral edema, and venous infarctions may occur. Additionally, acute respiratory distress may occur from nasal and laryngeal edema, necessitating urgent medical intervention [1].

SVC syndrome is a clinical diagnosis. The goal of imaging is to determine the etiology and anatomic location of the disease processes causing the patient's symptoms and to help formulate an appropriate treatment plan.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

A. Catheter venography upper extremity and SVC

Catheter venography upper extremity and SVC accurately delineates location and degree of obstruction and is helpful for planning interventions. However, it is not as useful for determining the cause of obstruction or assisting in staging evaluation in the case of obstruction caused by malignancy, and there is no relevant literature to support the use of catheter venography as an initial imaging evaluation, due to the invasive nature of the examination and associated risks, although it may be considered when rapid diagnosis and treatment is needed. CO₂ venography may be indicated in patients seeking an alternative imaging option. Zhao et al [5] demonstrated a high specificity (97%) but a low sensitivity (56%) for multidetector CT (MDCT) as compared with digital subtraction angiography (DSA). In their study, only 156 of 280 lesions found on DSA were correctly identified by MDCT. Therefore, catheter venography may be needed in addition to cross-sectional imaging for better characterization, particularly in catheter-related central venous stenosis in patients undergoing hemodialysis.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

B. CT chest with IV contrast

CT chest with intravenous (IV) contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. It allows delineation of degree and cause of SVC/brachiocephalic vein (BCV) obstruction/stenosis. CT chest with dynamic time-resolved component may be useful in the preoperative assessment of SVC syndrome to determine invasion into adjacent structures [6]. CT with IV contrast imaging, in general, has a high sensitivity (96%) and specificity (92%) for diagnosing SVC syndrome [7,8].

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

C. CT chest without and with IV contrast

CT chest without and with IV contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. It allows delineation of degree and cause of SVC/BCV obstruction/stenosis. CT chest without and with IV contrast will provide similar details as a CT chest with IV contrast. The addition of the CT chest without IV contrast is inferior for evaluation of the vasculature, although it can still add additional diagnostic value, particularly in cases in which there are central catheters or stents already in place.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

D. CT chest without IV contrast

There is no relevant literature to support the use of CT chest without IV contrast for the diagnosis of SVC syndrome. It is not indicated for acute presentation of SVC syndrome, as the vasculature is not adequately evaluated without IV contrast. It may be able to diagnose a chest/mediastinal mass or presence of a central venous catheter.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

E. CT neck and chest with IV contrast

CT neck and chest with IV contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. It allows delineation of degree and cause of SVC/BCV obstruction/stenosis. Although it may be able to provide additional information in cases in which jugular venous thrombus or obstruction is suspected, as the CT neck scan has a larger field of view, which includes the whole neck and the chest.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

F. CT neck and chest without and with IV contrast

CT neck and chest without and with IV contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. It allows delineation of degree and cause of SVC/BCV obstruction/stenosis. CT neck and chest without and with IV contrast will provide similar details as a CT neck and chest with IV contrast. It may be able to provide additional information in cases in which jugular venous thrombosis or obstruction is suspected, as the CT neck scan has a larger field of view, which includes the whole neck and the chest. The addition of the CT neck and chest without IV contrast is inferior for evaluation of the vasculature, although it can still add additional diagnostic value, particularly in cases in which there are central catheters or stents already in place.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

G. CT neck and chest without IV contrast

There is no relevant literature to support the use of CT neck and chest without IV contrast for the diagnosis of SVC syndrome, as the vasculature is not adequately evaluated without IV contrast. It may be able to diagnose a chest/mediastinal mass or presence of a central venous catheter.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

H. CTA and CTV chest with IV contrast

CT angiography (CTA) and CT venography (CTV) chest with IV contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. It allows delineation of degree and cause of SVC/BCV obstruction/stenosis. In most cases, it will provide additional or more accurate imaging of venous occlusion/thrombus due to the additional venographic phase (CTV). The arterial phase (CTA) may allow better characterization of chest/mediastinal tumors and possible pulmonary embolism (PE) if that is in the differential. Sundaram et al [9] suggest that CTV chest provides equivalent diagnostic information compared with CTA/CTV combination, with more streak artifact on the CTA phase.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

I. CTA chest with IV contrast

CTA chest with IV contrast in the arterial phase is a good initial imaging examination for most causes of SVC syndrome with acute presentation. However, because of the arterial timing of the contrast, it may limit delineation of degree and cause of SVC/BCV obstruction/stenosis compared with venographic phase (CTV). Therefore, it is inferior to CTA/CTV due to a lack of the venographic phase [9].

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

J. CTA chest without and with IV contrast

CTA chest without and with IV contrast in the arterial phase is a good initial imaging examination for most causes of SVC syndrome with acute presentation. However, because of the arterial timing of the contrast, it may limit delineation of degree and cause of SVC/BCV obstruction/stenosis compared with venographic phase (CTV). Therefore, it is inferior to CTA/CTV due to a lack of the venographic phase [9]. The without-IV contrast component (precontrast phase) is essentially a CT chest without IV contrast and can add additional diagnostic value, particularly in cases in which there are central catheters or stents already in place.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

K. CTV chest with IV contrast

CTV chest with IV contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. It allows delineation of degree and cause of SVC/BCV obstruction/stenosis. Lack of a CTA phase may be less useful to characterize chest/mediastinal tumors or diagnose PE if also suspected. Findings from Sundaram et al [9] suggest that CTV chest provides equivalent diagnostic information compared with CTA/CTV combination, with more streak artifact on the CTA phase.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

L. MRA and MRV chest without and with IV contrast

MR angiography (MRA) and MR venography (MRV) chest without and with IV contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. The venographic phase may allow better delineation of venous occlusion/thrombus. The image quality is highly dependent on equipment quality and protocol optimization. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

M. MRA chest with IV contrast

MRA chest with arterial IV contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. However, it may be less useful compared with an MRI/MRA/MRV combination if characterization of a mediastinal mass is indicated. The lack of an MRV may limit characterization of venous occlusion/thrombosis because the examination is optimized to evaluate the arteries. The image quality is highly dependent on equipment quality and protocol optimization. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

N. MRA chest without and with IV contrast

MRA chest without and with arterial IV contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. It may be less useful compared with an MRI/MRA/MRV combination if characterization of a mediastinal mass is indicated. The lack of an MRV may limit characterization of venous occlusion/thrombosis as the examination is optimized to evaluate the arteries. There is no advantage over MRA chest with IV contrast alone. The image quality is highly dependent on equipment quality and protocol optimization. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

O. MRA chest without IV contrast

MRA chest without IV contrast is not ideal as an initial imaging study; however, it may be considered in patients seeking an alternative option. The image quality is highly dependent on equipment quality and protocol optimization. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8]. However, it may be inferior to MRV chest for characterizing venous obstruction/thrombosis depending on protocol. It may also be less useful compared with an MRI/MRA/MRV combination if characterization of a mediastinal mass is indicated.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

P. MRI chest with IV contrast

MRI chest with IV contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. It may serve as a complementary imaging option to characterize a mediastinal mass more specifically. The image quality is highly dependent on equipment quality and protocol optimization. It is suboptimal for characterizing venous occlusion/thrombosis without MRA or MRV study concurrently. Metal artifacts may be a factor in image quality in patients with

stents/sternal wires/metal implants [8].

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

Q. MRI chest without and with IV contrast

MRI chest without and with IV contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. It may serve as a complementary imaging option to characterize a mediastinal mass more specifically, and the noncontrast portion of the study may provide additional diagnostic value compared with MRI chest with IV contrast alone. The image quality is highly dependent on equipment quality and protocol optimization. It is suboptimal for characterizing venous occlusion/thrombosis without MRA or MRV study concurrently. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

R. MRI chest without IV contrast

MRI chest without IV contrast may be considered in patients seeking an alternative option or as a complementary imaging option to characterize a mediastinal mass more specifically, although it may be inferior to other examinations using IV contrast. The image quality is highly dependent on equipment quality and protocol optimization. It is suboptimal for characterizing venous occlusion/thrombosis compared with all other MR studies. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

S. MRV chest without and with IV contrast

MRV chest without and with IV contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. It may be less useful compared with an MRI/MRA/MRV combination if characterization of a mediastinal mass is indicated or if PE is additionally suspected. However, it may be superior to MRA chest alone, with or without IV contrast, as MRV is optimized to evaluate venous structures. The image quality is highly dependent on equipment quality and protocol optimization. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

T. MRV chest without IV contrast

MRV chest without IV contrast is a good initial imaging examination for most causes of SVC syndrome with acute presentation. It may be less useful compared with an MRI/MRA/MRV combination if characterization of a mediastinal mass is indicated or if PE is additionally suspected. However, it may be superior to MRA chest with or without IV contrast because it is optimized to evaluate venous structures even without the additional benefit of IV contrast. Image quality is highly dependent on equipment quality and protocol optimization. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8]. This represents the only option, other than CO₂ venography and Doppler ultrasound (US), which does not involve the use of a contrast agent and provides superior cross-sectional information than the alternative modalities.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

U. Radiography chest

Radiography chest is not an optimal study for the diagnosis of SVC syndrome, although it may be able to diagnose a large chest or mediastinal mass, evidence of collaterals, or central catheter/stent [10,11]. However, it cannot determine extent of obstruction or cause of SVC syndrome.

Radiography often must be followed by a more definitive imaging study.

Variant 1: Adult. Suspected acute superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

V. US duplex Doppler upper extremity and chest

US duplex Doppler chest is not an optimal study for the diagnosis of SVC syndrome in the acute setting due to poor visualization of the mediastinum. However, it may be able to diagnose BCV or subclavian vein obstruction/thrombosis and may be able to visualize sequela of SVC obstruction/thrombosis, such as collaterals associated with central venous obstruction or slowed venous flow, or to diagnose upper extremity deep vein thrombosis (DVT) [10]. It cannot determine extent of obstruction or cause of SVC syndrome. If echocardiography is included in this category, it may be an initial study to diagnose pacemaker wire associated SVC syndrome [7].

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

Chronic SVC syndrome is a consequence of compromised venous drainage from the head, neck, and upper extremities, which typically presents over weeks, months, or years. The gradual development or worsening of a stenosis or occlusion allows time for collateral venous drainage to occur, decreasing symptom severity and acuity. The most common symptoms include swelling and discoloration of the face, neck, and upper extremities. This may be exacerbated by bending over or lying flat. Additional symptoms can include dyspnea, cough, dysphagia, dizziness, blurry vision, difficulty breathing, hoarseness, and cerebral edema [1,3].

SVC syndrome is a clinical diagnosis. The goal of imaging is to determine the etiology and anatomic location of the disease processes causing the patient's symptoms and to help formulate an appropriate treatment plan.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

A. Catheter venography upper extremity and SVC

Catheter venography upper extremity and SVC accurately delineates location and degree of obstruction and is helpful for planning interventions. However, it is not as useful for determining cause of obstruction and assisting in staging evaluation in the case of obstruction caused by malignancy, and there is no relevant literature to support the use of catheter venography as an initial imaging evaluation because of the invasive nature of the examination and associated risks. CO₂ venography may be indicated in patients seeking an alternative imaging option. Zhao et al [5] demonstrated a high specificity (97%) but a low sensitivity (56%) MDCT as compared with DSA. In their study, only 156 of 280 lesions found on DSA were correctly identified by MDCT. Therefore, catheter venography may be needed in addition to cross-sectional imaging for better characterization, particularly in catheter-related central venous stenosis in patients undergoing hemodialysis.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

B. CT chest with IV contrast

CT chest with IV contrast is a good initial imaging examination for most causes of chronic SVC syndrome. It allows delineation of degree and cause of SVC/BCV obstruction/stenosis. CT chest with dynamic time-resolved component may be useful in the preoperative assessment of SVC syndrome to determine invasion into adjacent structures [6]. CT with IV contrast imaging, in general, has a high sensitivity (96%) and specificity (92%) for diagnosing SVC syndrome [7,8].

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

C. CT chest without and with IV contrast

CT chest without and with IV contrast is a good initial imaging examination for most causes of chronic SVC syndrome. It allows delineation of degree and cause of SVC/BCV obstruction/stenosis. CT chest without and with IV contrast will provide similar details as a CT chest with IV contrast. The addition of the CT chest without IV contrast is inferior for evaluation of the vasculature, although it can still add additional diagnostic value, particularly in cases in which there are central catheters or stents already in place.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

D. CT chest without IV contrast

There is no relevant literature to support the use of CT chest without IV contrast for the diagnosis of SVC syndrome. It is not indicated for chronic SVC syndrome, as the vasculature is not adequately evaluated without IV contrast. It may be able to diagnose a chest/mediastinal mass or presence of a central venous catheter.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

E. CT neck and chest with IV contrast

CT neck and chest with IV contrast is a good initial imaging examination for most causes of chronic SVC syndrome. It allows delineation of degree and cause of SVC/BCV obstruction/stenosis. However, it may be able to provide additional information in cases in which jugular venous thrombus or obstruction is suspected, as the CT neck scan has a larger field of view that includes the whole neck and the chest.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

F. CT neck and chest without and with IV contrast

CT neck and chest without and with IV contrast is a good initial imaging examination for most causes of chronic SVC syndrome. It allows delineation of degree and cause of SVC/BCV obstruction/stenosis. CT neck and chest without and with IV contrast will provide similar details as a CT neck and chest with IV contrast, although the addition of the CT neck and chest without IV contrast is inferior for evaluation of the vasculature, although it can still add additional diagnostic value, particularly in cases in which there are central catheters or stents already in place. It may provide additional information compared with CT neck and chest with IV contrast if there are central catheters or stents in place. It may be able to provide additional information in cases in which jugular venous thrombosis or obstruction is suspected, as the CT neck scan has a larger field of view, which includes the whole neck and the chest.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

G. CT neck and chest without IV contrast

There is no relevant literature to support the use of CT neck and chest without IV contrast for the diagnosis of SVC syndrome, as the vasculature is not adequately evaluated without IV contrast. It may be able to diagnose a chest/mediastinal mass or presence of a central venous catheter.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

H. CTA and CTV chest with IV contrast

CTA and CTV chest with IV contrast is a good initial imaging examination for most causes of chronic SVC syndrome. It allows delineation of degree and cause of SVC/BCV obstruction/stenosis. In most cases, it will provide additional or more accurate imaging of venous occlusion/thrombus due to the additional venographic phase (CTV). The arterial phase (CTA) may allow better characterization of chest/mediastinal tumors and possible PE if that is in the differential. Sundaram et al [9] suggest that CTV chest provides equivalent diagnostic information compared with CTA/CTV combination, with more streak artifact on the CTA phase.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

I. CTA chest with IV contrast

CTA chest with IV contrast in the arterial phase is a good initial imaging examination for most causes of chronic SVC syndrome. However, because of the arterial timing of the contrast, it may limit delineation of degree and cause of SVC/BCV obstruction/stenosis compared with venographic phase (CTV). Therefore, it is inferior to CTA/CTV due to the lack of the venographic phase [9].

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

J. CTA chest without and with IV contrast

CTA chest without and with IV contrast in the arterial phase is a good initial imaging examination for most causes of chronic SVC syndrome. However, because of the arterial timing of the contrast, it may limit delineation of degree and cause of SVC/BCV obstruction/stenosis compared with venographic phase (CTV). Therefore, it is inferior to CTA/CTV due to the lack of the venographic phase [9]. The without IV contrast component (precontrast phase) is essentially a CT chest without IV contrast and can add additional diagnostic value, particularly in cases in which there are central catheters or stents already in place.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

K. CTV chest with IV contrast

CTV chest with IV contrast is a good initial imaging examination for most causes of chronic SVC syndrome. It allows delineation of degree and cause of SVC/BCV obstruction/stenosis. Lack of a CTA phase may be less useful to characterize chest/mediastinal tumors or diagnose PE if also suspected. Findings from Sundaram et al [9] suggest that CTV chest provides equivalent diagnostic information compared with CTA/CTV combination, with more streak artifact on the CTA phase.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

L. MRA and MRV chest without and with IV contrast

MRA and MRV chest without and with IV contrast is a good initial imaging examination for most

causes of chronic SVC syndrome. The venographic phase may allow better delineation of venous occlusion/thrombus. The image quality is highly dependent on equipment quality and protocol optimization. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

M. MRA chest with IV contrast

MRA chest with arterial IV contrast is a good initial imaging examination for most causes of chronic SVC syndrome. It may be less useful compared with an MRI/MRA/MRV combination if characterization of a mediastinal mass is indicated. The lack of an MRV may limit characterization of venous occlusion/thrombosis because the examination is optimized to evaluate the arteries. The image quality is highly dependent on equipment quality and protocol optimization. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

N. MRA chest without and with IV contrast

MRA chest without and with arterial IV contrast is a good initial imaging examination for most causes of chronic SVC syndrome. It may be less useful compared with an MRI/MRA/MRV combination if characterization of a mediastinal mass is indicated. The lack of an MRV may limit characterization of venous occlusion/thrombosis because the examination is optimized to evaluate the arteries. There is no advantage over MRA chest with IV contrast alone. The image quality is highly dependent on equipment quality and protocol optimization. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

O. MRA chest without IV contrast

MRA chest without IV contrast is not ideal as an initial imaging study; however, it may be considered in patients seeking an alternative option. The image quality is highly dependent on equipment quality and protocol optimization. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8]. However, it may be inferior to MRV chest for characterizing venous obstruction/thrombosis depending on the protocol. It may also be less useful compared with an MRI/MRA/MRV combination if characterization of a mediastinal mass is indicated.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

P. MRI chest with IV contrast

MRI chest with IV contrast is a good initial imaging examination for most causes of chronic SVC syndrome. It may serve as a complementary imaging option to characterize a mediastinal mass more specifically. The image quality is highly dependent on equipment quality and protocol optimization. It is suboptimal for characterizing venous occlusion/thrombosis without MRA or MRV study concurrently. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

Q. MRI chest without and with IV contrast

MRI chest without and with IV contrast is a good initial imaging examination for most causes of chronic SVC syndrome. It may serve as a complementary imaging option to characterize a mediastinal mass more specifically, as the noncontrasted portion of the study may provide additional diagnostic value compared with MRI chest with IV contrast alone. The image quality is highly dependent on equipment quality and protocol optimization. It is suboptimal for characterizing venous occlusion/thrombosis without MRA or MRV study concurrently. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

R. MRI chest without IV contrast

MRI chest without IV contrast is not ideal as an initial imaging study; however, it may be considered in patients seeking an alternative option or as a complementary imaging option to characterize a mediastinal mass more specifically, although it may be inferior to other examinations using IV contrast. The image quality is highly dependent on equipment quality and protocol optimization. It is suboptimal for characterizing venous occlusion/thrombosis compared with all other MR studies. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

S. MRV chest without and with IV contrast

MRV chest without and with IV contrast is a good initial imaging examination for most causes of chronic SVC syndrome. It may be less useful compared with an MRI/MRA/MRV combination if characterization of a mediastinal mass is indicated or if PE is additionally suspected. However, it may be superior to MRA chest alone, with or without IV contrast, as MRV is optimized to evaluate venous structures. The image quality is highly dependent on equipment quality and protocol optimization. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8].

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

T. MRV chest without IV contrast

MRV chest without IV contrast is a good initial imaging examination for most causes of chronic SVC syndrome. It may be less useful compared with an MRI/MRA/MRV combination if characterization of a mediastinal mass is indicated or if PE is additionally suspected. However, it may be superior to MRA chest with or without IV contrast, as it is optimized to evaluate venous structures even without the additional benefit of IV contrast. Image quality is highly dependent on equipment quality and protocol optimization. Metal artifacts may be a factor in image quality in patients with stents/sternal wires/metal implants [8]. This represents the only option, other than CO₂ venography and Doppler US, which does not involve the use of a contrast agent and provides superior cross-sectional information than the alternative modalities.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

U. Radiography chest

Radiography chest is not an optimal study for the diagnosis of SVS syndrome, although it may be

able to diagnose large chest or mediastinal mass, evidence of collaterals, or central catheter/stent [10,11]. However, it cannot determine extent of obstruction or cause of SVC syndrome. Radiography often must be followed by a more definitive imaging study.

Variant 2: Adult. Suspected chronic superior vena cava or brachiocephalic vein occlusion.

Initial Imaging.

V. US duplex Doppler upper extremity and chest

US duplex Doppler chest is not an optimal study for the diagnosis of SVC syndrome in the chronic setting due to poor visualization of the mediastinum. However, it may be able to diagnose BCV or subclavian vein obstruction/thrombosis and may be able to visualize sequela of SVC obstruction/thrombosis, such as collaterals associated with central venous obstruction or slowed venous flow, or to diagnose upper extremity DVT [10]. It cannot determine extent of obstruction or cause of SVC syndrome. If echocardiography is included in this category, it may be an initial study to diagnose pacemaker wire associated SVC syndrome [7].

Summary of Highlights

This is a summary of the key recommendations from the variant tables. Refer to the complete narrative document for more information.

- **Variants 1 and 2:** For initial evaluation of suspected acute or chronic SVC syndrome, contrast-enhanced chest CT scans, particularly CTA/CTV, with or without simultaneous inclusion of the neck are recommended studies. MRI with IV contrast and MRV/MRA chest with or without IV contrast are also recommended studies. The recommended CT and MR studies work well to diagnose and evaluate the cause and extent of SVC or BCV occlusion. MRI and CT should be used as alternative studies rather than complementary studies. On the other hand, CT or MRI scans without IV contrast, chest radiographs, US duplex Doppler, and catheter venography are usually not recommended due to their limited diagnostic value.

Supporting Documents

The evidence table, literature search, and appendix for this topic are available at <https://acsearch.acr.org/list>. The appendix includes the strength of evidence assessment and the final rating round tabulations for each recommendation.

For additional information on the Appropriateness Criteria methodology and other supporting documents, please go to the ACR website at <https://www.acr.org/Clinical-Resources/Clinical-Tools-and-Reference/Appropriateness-Criteria>.

Gender Equality and Inclusivity Clause

The ACR acknowledges the limitations in applying inclusive language when citing research studies that predates the use of the current understanding of language inclusive of diversity in sex, intersex, gender, and gender-diverse people. The data variables regarding sex and gender used in the cited literature will not be changed. However, this guideline will use the terminology and definitions as proposed by the National Institutes of Health.

Appropriateness Category Names and Definitions

| Appropriateness | Appropriateness | Appropriateness Category Definition |
|-----------------|-----------------|-------------------------------------|
|-----------------|-----------------|-------------------------------------|

| Category Name | Rating | |
|-----------------------------------|------------|--|
| Usually Appropriate | 7, 8, or 9 | The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients. |
| May Be Appropriate | 4, 5, or 6 | The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal. |
| May Be Appropriate (Disagreement) | 5 | The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel's recommendation. "May be appropriate" is the rating category and a rating of 5 is assigned. |
| Usually Not Appropriate | 1, 2, or 3 | The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable. |

Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, because of both organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared with those specified for adults (see Table below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® [Radiation Dose Assessment Introduction](#) document.

Relative Radiation Level Designations

| Relative Radiation Level* | Adult Effective Dose Estimate Range | Pediatric Effective Dose Estimate Range |
|---------------------------|-------------------------------------|---|
| 0 | 0 mSv | 0 mSv |
| | <0.1 mSv | <0.03 mSv |
| | 0.1-1 mSv | 0.03-0.3 mSv |
| | 1-10 mSv | 0.3-3 mSv |
| | 10-30 mSv | 3-10 mSv |
| | 30-100 mSv | 10-30 mSv |

*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (e.g., region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as "Varies."

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Disclaimer

The ACR Committee on Appropriateness Criteria and its expert panels have developed criteria for determining appropriate imaging examinations for diagnosis and treatment of specified medical condition(s). These criteria are intended to guide radiologists, radiation oncologists and referring physicians in making decisions regarding radiologic imaging and treatment. Generally, the complexity and severity of a patient's clinical condition should dictate the selection of appropriate imaging procedures or treatments. Only those examinations generally used for evaluation of the patient's condition are ranked. Other imaging studies necessary to evaluate other co-existent diseases or other medical consequences of this condition are not considered in this document. The availability of equipment or personnel may

influence the selection of appropriate imaging procedures or treatments. Imaging techniques classified as investigational by the FDA have not been considered in developing these criteria; however, study of new equipment and applications should be encouraged. The ultimate decision regarding the appropriateness of any specific radiologic examination or treatment must be made by the referring physician and radiologist in light of all the circumstances presented in an individual examination.

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