

Algorithm for percutaneous stenting in patients suffering from superior vena cava syndrome

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Background. Superior vena cava syndrome (SVCS) has been considered an emergent, life-threatening condition for a long time. The rate of causes of the syndrome has changed substantially since its first description by W. Hunter in a patient suffering from saccular aneurysm of syphilitic aorta in 1757.

Since the beginning of the era of radiotherapy, this was the main treatment modality for patients with SVCS. The emergent feature of the syndrome required immediate initiation of radiotherapy, often without the proper knowledge of the histopathological diagnosis of the SVCS underlying cause. The development of radiotherapy and chemotherapy in various types of cancer, and the development of supportive care in oncology and an understanding that SVCS is not an emergent life-threatening oncological condition evoked a need for treatment differentiation in SVCS patients.

Conclusions. Percutaneous stenting became a very efficient method in the treatment of SVCS patients since 1986. The purpose of stenting is supportive and/or palliative. The algorithm for stenting use has been developed. Algorithm is based on 4 questions emerging from daily clinical practice. To get valid responses, certain diagnostic procedures and tools are recommended and required.

1. Does the patient really suffer from SVCS?
2. What is the patient's general condition?
3. Is the stenting of SVCS contraindicated? (What is the origin of SVCS, what is the severity of SVCS?)
4. Is the histopathology of the process causing SVCS known? What is the histopathology of the process causing SVCS?

Responses to the questions above give reasons for the selection of a treatment modality. Rational usage of percutaneous implantable stents in properly chosen patients suffering from SVCS of malignant etiology ensures efficient differentiation of treatment modalities in supportive and/or palliative care of SVCS patients.

Key words: superior vena cava syndrome-therapy; stents; palliative care, algorithm for stenting

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Introduction

Superior vena cava syndrome (SVCS) is a critical condition diagnosed frequently by the symptoms and signs at the present time. SVCS may be caused by obstruction of SVC by tumorous invasion of venous walls and lumen or by extrinsic pressure of tumor mass both can be accompanied by intravascular thrombosis.^{1,2} SVCS was firstly reported and described by William Hunter in 1757.³ In the 17th century however, the majority of SVCS was caused by inflammatory conditions such as saccular aneurysm of syphilitic aorta as in Hunter's first case. Bronchogenic carcinomas and malignant lymphomas are responsible for a vast majority of SVCS today.⁴⁻¹¹ Moreover, there is a relatively new group of causes of SVCS that can be described as iatrogenic.¹¹ Proportions of malignant and benign causes of SVCS in the long run are given in Table 1. Recently, a comprehensive overview of SVCS published causes has been given elsewhere.¹¹

For a rather long period of time, SVCS has been suggested as a life threatening condition requiring an immediate treatment (usually radiotherapy) even without previous knowledge of tissue diagnosis.^{8,18-24} The development of radiotherapy tactics used in the treatment of SVCS was described in previous papers.^{8,18-20,24} Thus, radiotherapy became the main treatment modality for all (presumably) malignant cases of SVCS.^{1,25,26}

Experimental animal SVCS models, research of human SVCS causes, supportive care implementation and its results, better understanding of symptoms and signs development, revision of overall survival results were keystones of a change in the opinion on SVCS.^{4,27} Now, SVCS is not considered as an emergent, really life-threatening condition as originally thought of.^{4,9,28} Except in rare cases of SVCS with brain edema and/or intra-bronchial obstruction accompanied with significant dyspnoea, there is no reason to start with immediate treatment without proper tis-

sue diagnosis of the SCVS cause. The use of supportive treatment methods can diminish and alleviate symptoms and signs of SVCS for a time long enough to allow safe tissue biopsy and to establish the histopathological diagnosis of the SVCS cause.^{4,10,11,29}

The diagnosis is the most important for a more "specific" treatment of SVCS. Such treatment can ensure fast disappearance of symptoms and signs of SVCS and longer overall survival in certain cases in comparison with previously universally used radiotherapy.^{30,31}

Methods of supportive treatment in patients suffering from SVCS changed substantially. Intravenous stents started to be used in supportive and/or palliative treatment of SVCS in 1986.³² At present, several kinds of metallic stents are used.^{33,47} If properly indicated, stent indwelling may faster alleviate and exterminate the significant SVCS symptoms and signs.^{1,2,29}

The treatment with self-expandable or balloon-expandable stents is not suitable for all patients because there are contraindications in stent indwelling as well as in certain situations that can be debatable from the point of view of the achievable aim of treatment and cost-benefit ratio.^{42-44,48,49}

For the rational use of stents in supportive and/or palliative treatment of SVCS, the algorithm for stents indwelling has been developed. It is based on 4 questions that can be answered after the following examinations.²⁸

Question No. 1: Does the patient really suffer from SVCS?

Reason for the question No. 1

The question is necessary because several patients had been referred for SVCS treatment while they suffered from different diseases (like parotitis, dermatological diseases, allergic edema, status post tooth extraction).^{10,50}

Response

Response can be given by personal medical history, basic physical exam and duplex sonography assessing the blood flow of big veins. If the flow curve from duplex sonography is not physiological, computerized tomography with contrast medium and/or phlebography are necessary to obtain a more correct diagnosis.

If the response is "YES", go to the *Question No.2*.

If the response is "NO", go to the *Treatment No.1*.

Treatment No 1

Give different treatments for the specific (different) diseases.

Question No. 2: What is the patient's general condition?

Reason for the question No. 2

The question is needed because patients in very poor general condition are incapable to undergo diagnostic procedures. Their death can be imminent. The stenting would be not ethical in this situation.

Response

Response can be given by an evaluation of the patient's condition by the examination of his or her subjective and objective status (basic physical examination) and according to certain validated models. E.g.: Escalante's model,⁵¹ Karnofsky's scale etc. (Table 1).

If the response is "POOR", go to the *Treatment No. 2*.

Table 1. Escalante's validated model for predicting imminent death

<i>Progressive disease</i>
<i>Zubrod \geq 3</i>
<i>Triage Pulse \geq 110/min</i>
<i>Respiration \geq 28/min</i>

If the response is "GOOD", go to the *Question No. 3*.

Treatment No. 2

Palliative treatment at the hospice or at the palliative care unit for the symptomatic treatment (corticosteroids, diuretics, positioning, oxygen, antibiotics, etc.)

**Question No. 3: Is the stenting of SVC contraindicated?
(What is the origin of SVCS, what is the severity of SVCS?)**

Reason for the question No. 3

Stenting is contraindicated in patients with excessive obliteration of the large veins or their extensive invasion by of cancer (SVC and both brachiocephalic veins).

Table 2. Kishi's scoring system for signs and symptoms of SVCS obstruction

Signs and symptoms	grade
Neurological symptoms	
Stupor, coma, or blackout	4
Blurry vision, headache, dizziness, or amnesia	3
Changes in mentation	2
Uneasiness	1
Laryngopharyngeal or thoracic symptoms	
Orthopnea or laryngeal edema	3
Stridor, hoarseness, dysphagia, glossal edema, or shortness of breath	2
Cough or pleural effusions	1
Nasal and facial signs or symptoms	
Lip edema, nasal stiffness, epistaxis, or rhinorhea	2
Facial swelling	1
Venous dilatation	
Neck vein or arm vein distension, upper extremity swelling, or upper body plethora	1

Response

Response can be given by phlebography with digital subtraction (DSA) and spiral computerized tomography with contrast medium. Clinically, a severity of SVCS can be evaluated by Kishi's⁴⁸ or Nicholson's⁴⁹ classifications (Table 2).

If the response is "YES", go to the *Treatment No. 3*.

If the response is "NO", go to the *Questions No. 4*.

Treatment No. 3

Supportive care with corticosteroids, diuretics, oxygen, positioning, anticoagulants, antibiotics; relevant histopathology (endoscopy and/or biopsy); anticancer palliative therapy may be applied (chemotherapy or radiotherapy, or consider bypass).

Question No. 4a: Is the histopathology of the process causing SCVS known?

Reason for the questions No. 4

73 - 97% of SVCSs are caused by cancer. (Table 3)

The question is raised by different treatments of different cancers.

Table 3. The rate of benign and malignant causes of SVCS during the time since its first observation

Year	Author	Causes	
		Benign (%)	malignant (%)
1757	Hunter ³	97	3
1934	Ehrlich ¹²	54	46
1949	McIntire ¹³	67	23
1954	Schechter ¹⁴	40	60
1975	Lokich ⁸	3	97
1987	Fincher ¹⁵	13	87
1992	Baker ¹	10	90
1993	Escalante ²	3	97
1994	Tayade ¹⁶	13	87
1998	Kee ¹⁷	27	73

If the response is "NO", go to *Treatment No. 4A*.

If the response is "YES", go to *Question No. 4B*.

Treatment No 4A

Supportive care: Percutaneous stenting and endoscopy and/or bioptic methods to obtain tissue sample.

Question No. 4B: What is the histopathology of the process causing SVCS?

SVCS can appear in patients suffering from malignant tumors in three situations:^{10,44}

Group A: SVCS is the first sign of cancer (its histopathology is not known).

Group B: SVCS appears during a diagnostic process due to suspect for cancer (its histopathology can or cannot be known).

Group C: SVCS appears during the follow-up period (histopathology is usually known if cancer progression is revealed), in some cases, an oncological treatment is exhausted and cannot be repeated (group C1), in other cases, progressive disease is not proven - a cause of SVCS may be late sequels of the previous treatment (group C2).

According to our assessment of 151 patients with SVCS, the rates of patients in the groups are as follows: A - 25%, B - 65%, C - 10%.^{10,50}

Chemoresensitive cancers (groups A, B, C)

- Lymphomas
- Small cell lung cancer
- Germ cell tumors
- Etc

Rather chemoresistant cancers (groups A, B, C)

- Non small cell lung cancer
- Malignant melanoma

Progressive cancer - all oncological treatment is already exhausted (group C1)

- Further oncological treatment is not possible any more

The cause of SVCS is late sequel of previous oncological treatment (group C2)

- Radiation fibrosis of mediastinum

Treatment No. 4B

- Differentiated chemotherapy according to the diagnosis (as curative or palliative treatment of patients of groups A, B, and the rest of group C - see the group definitions)
- Percutaneous stenting or surgical methods (bypass) (both as a palliative treatment in patients of the groups C1 and C2 and as supportive care in patients of groups A and B and rest of group C), and
- Chemotherapy and/or radiotherapy (as curative or palliative treatment in patients of groups A, B, and the rest of group C)

In summary, endovascular treatment is a simple and safe procedure to restore the patency of SVC in patients with malignancies. In most cases, it should be indicated as the first-line treatment and performed as early as possible. With proper knowledge of the SVCS cases, the best results could be obtained, if stenting follows the correct treatment schedule.

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