

## Treatment of superior vena cava syndrome\*

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### ABSTRACT

The superior vena cava is formed by the union of the right and left brachiocephalic veins. It is located in the middle mediastinum, to the right of the aorta and anterior to the trachea. Superior vena cava syndrome consists of a group of signs (dilation of the veins in the neck, facial swelling, edema of the upper limbs, and cyanosis) and symptoms (headache, dyspnea, cough, orthopnea and dysphagia) caused by the obstruction of blood flow through the superior vena cava to the right atrium. This obstruction can be caused by extrinsic compression, tumor invasion or thrombosis. Such obstruction may also occur as a result of insufficient venous return secondary to intra-atrial or intraluminal diseases. From 73% to 93% of all cases of superior vena cava syndrome occur during the development of an intrathoracic tumor. Most patients presenting superior vena cava syndrome secondary to malignant neoplasms are treated without surgery, through radiotherapy, chemotherapy or the use of intraluminal stents. When the etiology of superior vena cava syndrome is benign, it can be treated with clinical measures (anticoagulation, raising the head, etc.) or, in refractory cases, with angioplasty, stents or surgery.

**Keywords:** Superior vena cava syndrome/therapy; Superior vena cava syndrome/surgery; Superior vena cava syndrome/etiology; Superior vena cava syndrome /drug therapy; Superior vena cava syndrome /radiotherapy; Superior vena cava

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## INTRODUCTION

Superior vena cava syndrome (SVCS) consists of a set of signs (dilation of the veins of the neck, facial plethora, edema of the upper limbs and cyanosis) and symptoms (headache, dyspnea, cough, orthopnea, dysphagia, etc.) resulting from obstruction of the blood flow through the superior vena cava to the right atrium (Tables 1 and 2).

The obstruction of the vena cava can be caused by extrinsic compression, tumor invasion, thrombosis or insufficient venous return secondary to intra-atrial or intraluminal diseases. Approximately 73% to 97% of SVCS cases occur during the evolution and expansion of intrathoracic tumors as a result of compression of the superior vena cava by the tumor itself or by the affected mediastinal lymph nodes.<sup>(1-2)</sup>

The type of cancer that most frequently causes SVCS (75% of all cases) is bronchogenic cancer, and 3% to 5% of patients with lung cancer develop SVCS over the course of the disease.<sup>(2-3)</sup> Lymphomas are the second leading neoplastic cause of the syndrome (15% of all cases), and 17% of lymphomas presenting mediastinal involvement result in SVCS.<sup>(4)</sup> Metastatic cancers account for 7% of all cases of SVCS.<sup>(5)</sup>

The physiopathology of SVCS was reviewed, demonstrating that the superior vena cava is vulnerable to obstruction caused by the following factors: its strategic location in the visceral compartment of the mediastinum, surrounded by rigid structures (such as the sternum, trachea, right mainstem bronchus, aorta and right pulmonary artery); its thin, easily compressed walls; the transport of blood at low pressures; and the fact

that it is completely circumscribed by the mediastinal (subcarinal, perihilar and paratracheal) lymph nodes. These factors can explain many of the clinical and pathological aspects of the syndrome.<sup>(6)</sup>

A great number of collateral blood vessels are recruited when the vena cava and its principal venous tributaries become obstructed. In this circumstance, collateralization occurs via extracavitary venous networks, principally in the skin and musculature of the chest wall. In addition to the obstruction, the high venous pressure also led to the appearance of shunts in the veins and adjacent low-pressure plexuses. Over the course of weeks or months, this consistently elevated pressure leads to progressive distention and dilation of the collateral veins, which can increase in caliber, thereby increasing blood flow.

## TREATMENT OPTIONS

The treatment of SVCS depends on the severity of the symptoms and the cause of the obstruction, as well as on the histological type and stage of the tumor produced. Measures such as reclining the patient and elevating the head, as well as controlling the volume of oxygen administered and the oxygen supplementation, are valid alternatives for use prior to diagnosis and initiation of the definitive treatment. The roles played by diuretics and corticosteroids in the treatment of SVCS remain unclear. Despite being described as a medical emergency, SVCS rarely acquires that characteristic. In a review of 90 studies describing 1,986 cases de SVCS, only one death, from bronchial aspiration of epistaxis, was directly attributed to SVCS.<sup>(7)</sup> Among the serious complications described are cerebral and laryngeal edema, although these are rarely seen in

TABLE 1

Superior vena cava syndrome: symptoms<sup>(10)</sup>

Symptom	Frequency
Suffusion	80%
Dyspnea	63%
Cough	55%
Pain	20%
Dysphagia	12%
Syncope	7%
Edema of the upper limbs	3%
Orthopnea	2%
Obnubilation	2%
Lethargy	1%
Stridor	1%

TABLE 2

Superior vena cava syndrome: physical examination<sup>(10)</sup>

Findings in the physical examination	Frequency
Distention of the thoracic veins	67%
Facial edema	60%
Distention of the veins of the neck	58%
Labored exhalation	50%
Facial plethora	20%
Edema of the upper limbs	14%
Cyanosis	13%
Paralysis of the vocal cords	3%
Horner's Syndrome	2%

current practice. In fact, cerebral edema is often related to metastatic foci in the brain and respiratory impairment (consequent to compression of the trachea by the tumor) and not as a result of the SVCS per se. Therefore, as previously mentioned, the treatment of SVCS can be initiated prior to an appropriate evaluation of its etiology.<sup>(8-9)</sup>

The majority of the patients with SVCS secondary to malignant neoplasms can be treated without surgery, through radiotherapy or chemotherapy or through placement of intraluminal stents. From 46% to 70% of patients with bronchogenic carcinoma respond to radiotherapy (or to chemotherapy and radiotherapy combined) and experience relief from their symptoms within the first two weeks of treatment. This improvement can be attributed to vena cava permeability being re-established or to the enhancement of the collateral veins. In general, the reduced venous distention and subjective improvement of symptoms do not occur until three to seven days after the initiation of treatment.<sup>(10)</sup>

## RADIOTHERAPY

The use of radiotherapy in patients with SVCS prior to receiving the histological diagnosis is currently considered inappropriate. The presence of SVCS does not impede the adoption of appropriate curative treatment when possible. Before radiotherapy is started, general therapeutic measures, such as raising the head of the bed and administering corticosteroids and diuretics, can be taken. In patients with SVCS and small cell lung cancer, although radiotherapy has been used in some studies, the most appropriate therapeutic approach is combined chemotherapy. In such cases, there are no differences between the two approaches in terms of the time to resolution or in terms of results. However, chemotherapy offers the advantage of treating the disease systemically, as well as that of avoiding high radiation loads on the heart and lungs.<sup>(11)</sup> In 43% to 100% of cases, resolution of the syndrome is achieved in seven to ten days.<sup>(12-13)</sup> In a randomized study involving patients with SVCS and small cell lung cancer, the value of radiotherapy was evaluated in relation to chemotherapy in patients initially treated with chemotherapy, and it was found that such patients gained no benefit from radiotherapy.<sup>(14)</sup> Nevertheless, in patients with SVCS and non-small cell lung cancer, radiotherapy plays

a central role. Based on somewhat limited evidence of a more rapid response,<sup>(10)</sup> initial treatment with two to four fractions of 300 to 400 Gy has been recommended.<sup>(11)</sup> However, the timing, dose and fractioning of the radiotherapy applications for SVCS have not been definitively established, and there is no evidence indicating the size of the final dose required to obtain the best clinical response. In general terms, in non-small cell lung cancer, the total dose used is 60 Gy, whereas doses of 20 to 40 Gy have been used in lymphomas and neoplasms that are more radiosensitive. Whenever possible, all locoregional disease, including that found in the hilar and supraclavicular regions, with appropriate margins, should be treated. It is of note that the doses used in radiotherapy can vary not only depending on the histological nature of the tumor but also on whether the radiotherapy was or was not combined with chemotherapy and whether the therapeutic objective was palliative or curative.<sup>(15)</sup>

The majority of patients treated with a course of radiotherapy respond to the treatment and experience relief of symptoms within a matter of days. In one study, it was reported that a subjective response to radiotherapy was achieved within seven days after the initiation of treatment in 91% of the cases.<sup>(12)</sup> The authors observed an objective response after fourteen days in 89% of the patients studied. In another study, more rapid symptom relief was achieved with high radiotherapy fractions, 70% of the patients presenting a response in two weeks or less, compared with 56% of the patients treated with conventional radiotherapy fractions.<sup>(10)</sup> In the same study, patients with lymphoma presented a better response to treatment than did those with bronchogenic carcinoma. Failure to obtain symptom relief with radiotherapy was seen in 13% of cases. This treatment failure appeared to be related to thrombi in the superior vena cava. The better survival of patients treated with radiotherapy runs parallel to the relief of symptoms and has been correlated with tumor histology.<sup>(10)</sup> Patients with lymphoma presented a five year survival rate of 41%, whereas patients with small cell carcinoma present a one year survival rate of 24% and a five year survival rate of 5%. However, patients with other types of bronchogenic carcinoma present a 17% survival rate, dropping to 2% over two years.<sup>(10)</sup> These data demonstrate that prolonged survival, and possibly the cure of the disease, can be attained through

the use of radiotherapy.

Failure to resolve SVCS should always raise the suspicion of accompanying complications such as thrombi in the superior vena cava. When SVCS is recurrent or refractory to treatment, especially in patients having received radiotherapy, the placement of intravascular stents might be required in order to re-establish vena cava permeability.<sup>(16)</sup>

## CHEMOTHERAPY

In the treatment of SVCS secondary to chemosensitive cancers such as lymphoma or small cell lung cancer, chemotherapy can be effective either as a primary treatment or in combination with radiotherapy. For chemotherapy to be considered, it is fundamental that a histological diagnosis be made. In recent decades, the development of effective drug combinations has allowed chemotherapy to be used as the treatment of choice in SVCS consequent to small cell lung cancer. In one study, seven patients with small cell carcinoma were treated with chemotherapy (Iomustine, cyclophosphamide and methotrexate), and immediate resolution of the signs and symptoms of the syndrome was observed.<sup>(17)</sup> Similar results were obtained by another group of authors studying a sample of 22 patients treated with combined chemotherapy.<sup>(12)</sup> Complete resolution of the syndrome was observed in 21 of those patients within fourteen days after the initiation of the treatment. In one study, the history of SVCS from small cell lung cancer at the M.D. Anderson Hospital was reviewed.<sup>(13)</sup> It was found that 18 patients were treated with radiotherapy alone, 18 were treated with systemic chemotherapy alone, and 7 were treated with a combination of chemotherapy and radiotherapy. All of the therapeutic modalities were rapidly effective in relieving of the symptoms of superior vena cava obstruction. However, the use of chemotherapy alone was associated with a greater proportion of premature deaths.

Chemotherapy can also be used as an initial treatment for SVCS from lymphoma or from other highly chemosensitive cancers. In a review of 30 cases of SVCS due to lymphoma, it was observed that 8 patients received only radiotherapy, received only chemotherapy (various different regimens), and 12 were treated with a combination of the two.<sup>(4)</sup> The results of that study demonstrate that, by two

weeks after the initiation of the treatment, chemotherapy and radiotherapy were equally effective in the relief of the symptoms of the syndrome. As expected, chemotherapy alone or chemotherapy in combination with radiotherapy was superior to radiotherapy alone in terms of overall survival as well as in terms of disease-free survival. However, including radiotherapy resulted in fewer local recurrences. The authors concluded that the initial treatment of patients with SVCS secondary to lymphomas aimed at both the systemic and localized disease, recommending that all patients receive systemic chemotherapy. The authors also recommend that, when the tumor is larger than 10 cm in horizontal diameter and the histological diagnosis indicates large cell lymphoma, the chemotherapy be followed by local irradiation of the mediastinum. Such chemotherapy can be highly effective and presents an alternative to radiotherapy in the initial approach to SVCS due to large cell lymphoma or lung cancer.

## EXPANDABLE INTRALUMINAL DEVICES - STENTS

Intravascular stents are devices that can be placed in the lumen of a blood vessel and, after being expanded, support the walls of the vessel, counteracting the intrinsic and extrinsic collapsing forces. Innumerable stent designs are currently being evaluated. Intravascular stents can be classified by their mechanism of expansion (auto-expandable, thermo-expandable or balloon-expandable) or by other properties such as absorbability recoverability and coatings. The auto-expandable stents are compressed within an application catheter, inserted into an artery or vein and expand to a predetermined diameter when the catheter is withdrawn, being held in position by an embolus. Such stents are highly flexible and are relatively easy to apply, although they might not resist the radial compression exerted by the vessel wall. Many systems of auto-expandable stent introduction are of a smaller diameter than those used to introduce balloon-expandable stents, thereby reducing complications at the puncture site. Balloon-expandable stents are compressed within an angioplasty balloon prior to insertion into the vessel. When in the desired position, the balloon is inflated and the stent is expanded. Many

balloon-expandable stents have relatively good tensile strength but can lack longitudinal flexibility. Despite the fact that the rigidity of these stents makes them difficult to place within tortuous blood vessels, this same characteristic can be advantageous in creating a stable, stationary surface, which facilitates early re-endothelization. When necessary, balloon-expandable stents can be re-inflated with a larger balloon. Such recuperation is not possible with auto-expandable or thermo-expandable stents. Finally, one group of researchers developed and described the prototype of a thermo-expandable stent composed of a titanium alloy designated nitinol. This particular stent also has the unparalleled quality of thermal contraction. When heated, the stent can be rectified and will maintain this form until cooling to below the ambient temperature.<sup>(18)</sup> Upon subsequent insertion into a blood vessel and therefore exposed to body temperature, the guide "remembers" its original form and returns to its spiral shape.<sup>(19)</sup>

In recent years, the use of intraluminal stents has become an important alternative treatment for SVCS of various etiologies, allowing rapid relief of symptoms without the need for major surgery, while the patient continues receiving specific treatment for the disease responsible for the syndrome.<sup>(20)</sup>

## TYPES OF STENTS USED IN THE TREATMENT OF SVCS

There are various models of stents that can be used in the treatment of SVCS. Since the superior vena cava is a blood vessel of a large caliber, the stent must also be of a large diameter, in the majority of the cases, from 12 to 14 mm.

The Gianturco stent was the first stent used in the treatment of SVCS. It is an auto-expandable stent composed of stainless steel and presenting a zigzag configuration in a rigid cylindrical form. Its expansion force depends on various factors, such as caliber and diameter, as well as quantity and form of its angles. The expansion force of the stent increases in parallel with shorter length, greater diameter and greater angle achieved by its curvature. The recommended stent diameter is 1.25 to 1.5 times the diameter of the vessel. Catheters used to introduce a stent should therefore have a diameter of 8 to 16 F.

The Wallstent stent is an auto-expandable stent composed of stainless steel filaments in a tubular configuration. Catheters from 7 to 9 F are used to implant Wallstents. The Wallstents available for use in SVCS vary in diameter from 10 to 24 mm, the 16-mm stent being the most widely used. Its greater flexibility allows it to easily conform to the curvature of blood vessels. The length of the stent reduces by 30% when it is completely expanded, which makes precise positioning difficult.

The Palmaz stent is balloon-expandable and is contained within a thin-walled stainless steel tube. Experimental studies of metallic stents in animals have demonstrated complete endothelization of such stents within approximately four weeks after its intravascular implantation. The lumens of the vena cava and of its collateral branches remain permeable after the implantation of a stent.<sup>(21)</sup> The diameter of the stent should not be expanded by more than 20% since this could increase the risk of acute thrombosis and pronounced intimal hyperplasia.

## Indications and contraindications

Despite the fact that the primary treatment of SVCS in patients with malignancies is radiotherapy or, in some cases, chemotherapy, the placement of stents should be considered when conventional treatment fails. The use of stents should be seen as an important therapeutic alternative since approximately half of all patients who initially respond to radiotherapy present recurrence of the symptoms.<sup>(22)</sup> Angioplasty and stent placement should also be considered in patients presenting severe acute symptoms requiring immediate treatment since radiotherapy and chemotherapy might not relieve symptoms promptly.<sup>(23)</sup>

Stent placement should be considered for any patient presenting SVCS secondary to a malignant tumor. In this population of patients, life expectancy is only three to ten months, and the objective of the treatment is purely palliative, aimed at relieving symptoms and improving quality of life as rapidly as possible.<sup>(24)</sup>

Some authors consider the invasion of the superior vena cava by a tumor to be a contraindication to stent placement, given the possibility that the tumor might grow through the interstices of the stent.<sup>(23)</sup> However, some studies have reported that the use of metallic stents in

patients with SVCS secondary to invasive tumors provides good results.<sup>(20)</sup> In early studies, occlusion of the superior vena cava was considered a contraindication to stent placement. Many later studies demonstrated recanalization of the occluded segment of the superior vena cava after appropriate stent placement, achieving immediate relief of symptoms that are often incapacitating.<sup>(20)</sup>

The use of stents in the treatment of SVCS secondary to benign processes is controversial. In such cases, the disease occurs in relatively young patients, whose life expectancy is virtually unaltered by the disease. Therefore, the result of the treatment in these cases must be long-lasting. Currently, the initial treatment of such patients includes anticoagulation and angioplasty. If the initial treatment fails, stent placement, which will not hinder future surgical interventions, can be considered.<sup>(20)</sup>

#### Technical considerations

Before a stent is placed into the superior vena cava, a venogram, in two different positions, should be performed in order to determine the extent, severity and location of the obstruction. Prior to the intervention, the collateral venous network should be carefully evaluated, and any thrombi or tumor invasions should be investigated. The venographic classification of the obstruction, according to the criteria established by Stanford and Doty, should also be determined in order to estimate the possibility of complications such as cerebral edema and respiratory impairment (Chart 1).<sup>(25)</sup> The measurement of pressures can also aid in the determination of the SVCS severity. In one study sample, the placement of stents was only indicated when the peripheral pressure of the vena cava was greater than 22 mmHg.<sup>(26)</sup> Of the 9 SVCS patients in that sample, 3 presented pressure lower than 22 mmHg and were therefore not submitted to stent placement. In those cases, the syndrome remained stable, without therapeutic intervention, up until the death of the patients from the underlying disease. However, the majority of the authors have based their therapeutic decisions solely upon clinical findings.

The most common percutaneous approach to stent placement in the superior vena cava is via the right common femoral vein. In cases of superior vena cava occlusion or highly pronounced stenosis,

stent placement can be achieved through more than one access, such as via the right and left external jugular veins, or via the peripheral veins of the arm.

#### Clinical results

Since the first studies reporting the success of stent placement in the treatment of SVCS, numerous additional studies have been carried out, in which complete resolution of the syndrome was reported in 68% to 100% of the cases.<sup>(27-32)</sup> Regardless of the type of stent used, immediate relief of symptoms such as headache has been reported by various authors. Cyanosis and facial edema improve in one or two days,<sup>(31-34)</sup> and edema of the upper limbs is generally resolved two or three days after the insertion of the stent<sup>(23,30,34)</sup> but can persist for up to one week.<sup>(16,26)</sup>

In a study published in 1987, one group of authors related their experience with expandable Gianturco metallic stents in the treatment of two patients suffering recurrence of the syndrome after radiotherapy. The authors observed immediate relief of the obstructive symptoms, as well as a favorable palliative result in the short-term (six months), in both patients.<sup>(31)</sup> In fact, prior to that study, the Gianturco stent had been used in the treatment of SVCS by other authors (in 1986). However, in that previous study, the patient died (as a result of chemotherapy toxicity) three weeks after stent placement. Nevertheless, the vein was found to be permeable during the autopsy.

Despite the fact that the life expectancy of patients with SVCS is relatively short, the majority of the stents used in research remain permeable throughout the remainder of the life of the patients. In a study of thirteen patients submitted to stent

Chart 1 - Venographic classification

Category	Description
I	Up to 90% stenosis in the vena cava superior with permeable azygos vein
II	More than 90% stenosis in the vena cava superior with permeable azygos vein and flow toward the right atrium
III	More than 90% stenosis in the vena cava superior with reverse flow in the azygos vein
IV	Complete obstruction of the vena cava superior and of one or more of its major tributaries



placement and monitored for three days to six months, no recurrence was observed.<sup>(36)</sup> In another study, involving eleven patients monitored for one to eleven months, only one case of recurrence was observed.<sup>(37)</sup> The authors of yet another study achieved clinical success in 93% of the patients treated, with no recurrences during a one- to fourteen-month follow-up period.<sup>(23)</sup> In still another study, no long-term follow-up evaluation was made because the patients all died prematurely from the underlying disease. However, as a palliative treatment, the authors found stent placement to be successful in 55% of the cases.<sup>(38)</sup> The rates of recurrence reported in the literature range from 0% to 45%.<sup>(36)</sup> The recurrence of the obstruction of the superior vena cava, secondary to the growth of the tumor in the interstices or along the stent borders and resulting in thrombosis, can be related to various factors. In three different studies involving a total of 56 patients, one case of recurrence of SVCS secondary to the growth of the tumor was reported.<sup>(28-29,37)</sup> The authors of all three studies employed Gianturco stents, suggesting that the open architecture of the stent leaves it more susceptible to proliferation of the tumor. Such tumor infiltration has not been reported with use of the Wallstent or Palmaz stents, although tumor growth can occur along the borders of all types of stents, resulting in recurrence of the symptoms.

Thrombosis of the superior vena cava or of the brachiocephalic vein after stent placement is not uncommon. In two different studies, thrombosis occurred (one case in each study) soon after the placement of the stent in the superior vena cava in patients who had not been submitted to anticoagulation.<sup>(29,31)</sup>

The recurrence of SVCS can be treated using interventional radiological techniques. Thrombolysis, angioplasty, and the placement of an additional stent have been reportedly used as successful treatment modalities in cases of recurrence.<sup>(32,39)</sup> In one study, it was reported that five SVCS patients treated with stent placement developed recurrence of the syndrome.<sup>(38)</sup> All five patients were submitted to re-treatment with thrombolysis or placement of an additional stent and remained asymptomatic up until their death.

### Complications

Complications related to stent placement are

infrequent. In one study, SVCS patients were treated with a combination of stent placement and thrombolysis, and a 10% rate of complications was observed.<sup>(40)</sup> Migration of the stent is a relatively rare complication, inappropriate placement being a determinant of its occurrence.<sup>(34)</sup> Migrating stents can follow the blood flow to the heart and lung, causing arrhythmias and other complications, including death. In one study, it was reported that a Gianturco stent migrated to the right atrium five weeks after its placement.<sup>(28)</sup> In another study, a Palmaz stent was observed to migrate to the left pulmonary artery, being subsequently removed through percutaneous radiological techniques.<sup>(40)</sup> To reduce the risk of migration, some authors recommend the incomplete dilation of the stenosis prior to the placement of the stent.<sup>(34)</sup> This allows the device to adequately adapt at the site of the stenosis before being fully expanded with a balloon catheter of the appropriate size.

Normalization of venous return after stent placement can cause cardiac insufficiency. Pulmonary edema was described in a patient presenting vena cava pressure of 22 mmHg at the periphery of the stenosis, dropping to 9.6 mmHg after the placement of the stent. Mean atrial pressure increased from 8.1 mmHg to 9.6 mmHg. Pulmonary edema, diagnosed two hours after the placement of the stent, was resolved through clinical treatment.<sup>(26)</sup> One death from respiratory failure at five hours after the placement of the stent has been described in a patient with altered pulmonary function who developed acute cor pulmonale.<sup>(40)</sup>

### Surgical treatment

In view of the favorable results obtained with chemotherapy and radiotherapy, surgical treatment is rarely necessary in SVCS. Two-thirds of all SVCS patients present symptom relief within one to two weeks with nonsurgical treatment. Many authors believe that performing vascular graft-type bypasses has no place in the treatment of SVCS secondary to malignant diseases,<sup>(41)</sup> whereas others admit that this therapy can be used in a highly select patient population.<sup>(9)</sup> The advantage of surgery is the immediate and sustained relief of the symptoms, together with the immediate resolution of vena cava obstruction. The disadvantages of surgery include the morbidity and

mortality associated with the surgical procedure, principally due to the subjacent neoplastic process. In addition, it has been observed that these patients present an increased risk of bleeding consequent to the distention of the veins in the upper compartment.<sup>(42)</sup> Despite these drawbacks, surgery can play a well-defined, albeit limited, role in the treatment of SVCS.

The possible indications for surgical intervention described in the literature include neoplasms that are refractory to treatment (radiotherapy and chemotherapy) and thrombi in the superior vena cava or in its major tributaries, as well as acute occlusion of the superior vena cava accompanied by severe symptoms.<sup>(43)</sup> Another indication for surgery is the recurrence of SVCS after a complete cycle of radiotherapy and chemotherapy.<sup>(44)</sup> Finally, surgical intervention can also be beneficial for patients with benign processes obstructing the superior vena cava. In addition to being a treatment for the SVCS, such interventions also allow the collection of tissue for use in making a definitive histological diagnosis. More recently, however, such relative indications for surgery have been re-evaluated in view of the development of less invasive percutaneous techniques, such as angioplasty and placement of intraluminal stents. These new techniques provide immediate relief of symptoms, together with lower morbidity and mortality than that presented by the surgical procedure.<sup>(20)</sup> Therefore, the majority of recent studies of surgical treatment of SVCS include patients with benign diseases (fibrosing mediastinitis, thrombosis caused by catheters or pacemaker electrodes, or spontaneous thrombosis of the superior vena cava).<sup>(45)</sup> Nevertheless, surgical intervention has been used successfully for relief of SVCS due to malignant neoplasms.<sup>(44,46-47)</sup>

### Surgical procedures

Many procedures have been described for the treatment of SVCS secondary to malignant or benign processes. There are two basic categories of surgical treatment: resection and bypass. Bypass procedures create a new course along which blood flows to the right atrium, avoiding the obstructed segment of the vena cava. The tumor is not addressed. A synthetic graft or simply a large caliber vein (jugular, brachiocephalic or subclavian) is used to achieve the bypass. The other technique

involves en bloc resection of the superior vena cava and of the tumor, followed by reconstruction of the vena cava and interposition of a graft. Thrombectomy can be combined with surgery in various circumstances (some degree of thrombosis accompanies 20% of cases). Thrombotic occlusion of the brachiocephalic, subclavian or jugular vein can impede the execution of the bypass.

Various bypass techniques for diverting the superior vena cava have been used in recent years. These techniques can be divided into two major groups: grafts made of synthetic materials; and autologous vein grafts. The distal end of the anastomosis can be at a number of sites, including the femoral vein, the azygos vein, the vena cava inferior, and the auricle of the right atrium. The auricle of the right atrium has been described as the site most frequently used and presenting the best success rate, probably due to its easy access and appropriate size.<sup>(46)</sup> There are some prerequisites for the use of a material as a graft: having a nonthrombogenic surface; being sufficiently rigid to resist external compression and constriction of the suture line; presenting internal pressure and flow high enough to maintain graft permeability. A Dacron graft between the left brachiocephalic vein and the auricle of the right atrium was used successfully in four cases of SVCS caused by bronchial carcinoma.<sup>(48)</sup> Good results were obtained for a period of five to fourteen months, although partial obstruction was observed in one of the grafts at seven weeks after the procedure. Various autologous vein grafts have been described and used with success. A graft using the right accessory saphenous vein was described in an SVCS patient presenting recurrence of the symptoms seven months after palliative radiotherapy.<sup>(49)</sup> This technique uses the autologous saphenous vein to construct the anastomosis via a subcutaneous tunnel and using an accessory vein. Ideally, however, the superior vena cava requires a conduit of a diameter similar to its own. In one study, autogenic femoral vein segments were used to create the grafts implanted in five patients.<sup>(50)</sup> However, a bypass technique that has been widely used of late is that of "spiral" autogenic vein graft first described by Doty<sup>(51)</sup> in 1982 and later adapted by Smith & Brantigan,<sup>(47)</sup> as well as by others. Doty used this technique with success to divert the superior vena cava in five patients with fibrosing



mediastinitis and in six patients with bronchogenic carcinoma.<sup>(51)</sup> All of the patients experienced relief of the SVCS symptoms, without significant postoperative morbidity. In patients presenting obstruction due to benign disease, long-term relief of symptoms was achieved, with a follow-up period of three months to six years. Satisfactory improvement was obtained for twelve months or more in patients presenting malignancies. Similar results haqdo for mudar eu

The results obtained through surgery in these studies, although they were highly selective and limited, lend support to the recommendation that this treatment alternative be used under circumstances in which traditional therapeutic interventions have been inefficacious (recurrent obstruction after palliative chemotherapy or extensive venous thrombosis), or even in certain symptomatic cases of SVCS caused by benign diseases.

When a vein segment of an appropriate length and diameter is available, this should be considered the material of choice for the graft. The femoral vein is rarely used due to the possibility that edema of the upper limbs will occur after its removal. One alternative that merits mention is construction of a spiral graft from a long autogenic segment of saphenous vein.<sup>(53)</sup> Doty & Baker were the first to apply this technique.<sup>(51)</sup> Doty reported using spiral vein grafts as the primary treatment in ten patients with total occlusion of the superior vena cava.<sup>(46)</sup> The graft was used to connect the brachiocephalic artery (or the left internal jugular vein) to the right atrium, with an extension ranging from 9 to 13 mm. All of the patients presented relief of the signs and symptoms 48 hours after the operation, all of the grafts proved permeable (radiographically) at seven days to eighteen months after the operation, and all of the patients with benign disease were found to be in a good clinical state after three months to six years of follow-up evaluation. Doty et al. currently assert that, at least in benign diseases, the spiral composition of the vein graft constitutes an excellent superior vena cava graft.<sup>(54)</sup> In their fifteen years of experience with vein grafts used for bypass in nine patients, seven of the grafts remained permeable for more than fifteen years. The selection of patients for the procedure was based on the venographic classification previously described.<sup>(24)</sup> Symptomatic patients presenting

complete obstruction of the superior vena cava and formation of an extensive collateral network were chosen, and the venogram allowed the most appropriate point for the anastomosis to be identified.

Together with the autologous grafts, polytetrafluoroethylene grafts constitute an alternative that has been in clinical use for a sufficiently length of time to be recommended for use as a bypass or as a substitute for the superior vena cava. Some authors<sup>(9)</sup> have used polytetrafluoroethylene grafts in patients with various malignant processes. The authors observed short-term permeability of the graft in twelve of the thirteen patients studied. Since only 27% of the patients survived for three years, a long-term evaluation could not be carried out. Other authors have also had success in substituting the superior vena cava with polytetrafluoroethylene grafts in patients with stage III thymoma.<sup>(55)</sup>

Patients with invasive thymoma and presenting vascular infiltration or other benign tumors involving or circumscribing the vena cava, but without obstruction or thrombosis of the blood vessel, are candidates for excision and replacement of the vein. This possibility can be anticipated in the preoperative period through tomographic evaluation. Complete resection of the lesion is mandatory if the resection of the vein is under consideration.

For substitution of the superior vena cava with a polytetrafluoroethylene graft of 18 to 20 mm in diameter is generally necessary. If the proximal anastomosis site is the brachiocephalic vein, a graft of 10 to 14 mm in diameter should be used. In general, one of the brachiocephalic veins is used in the reconstruction. A "Y" graft can be performed, but graft-graft anastomoses should be avoided. The blood vessel is clamped, and the mass is excised en bloc. Clamping of the vena cava is well tolerated for up to an hour. The distal and proximal anastomoses are performed with a continuous suture, thus avoiding graft kinking.

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