A sword threatening the heart: The scimitar syndrome

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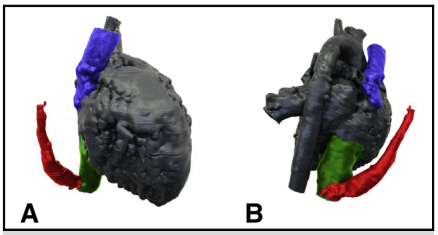
Video clip is available online.

Scimitar syndrome (SS) is a rare congenital heart anomaly consisting of anomalous venous drainage of part or the entire right lung into the upper portion of the inferior vena cava (IVC), right lung hypoplasia, and a variable systemic arterial blood supply to the right lung.[1,2]

This malformation has been described by many pathologists over time. In an article published in 1956 by Halasz and colleagues,[3] the word scimitar was used for the first time to describe the shape of this anomalous pulmonary venous connection. However, it was only in 1960 that Catherine Neill and colleagues[4] named the syndrome, characterizing it and linking it to the radiologic appearance of a sword adjacent to the edge of the right heart that can sometimes be found at diagnosis.

The incidence of SS is relatively low, representing 3% to 6% of the partial anomalous venous connections.[1] Previous studies were based on single-center experiences or rather small, multicenter studies.[5-13] However, our group was recently able to conduct a multicenter study on behalf of the European Pediatric Cardiology Association and the European Congenital Heart Surgeons Association.[14] This led to the collection of nearly 500 patients with SS from 51 centers, the largest series of patients with SS evaluated so far. In this review, we arbitrarily divided patients into 3 groups according to their age at diagnosis: neonates/infants (<1 year), children (between 1 and 10 years), and adolescents/adults (Table 1).



Three-dimensional silicon-printed model of the heart of a patient with scimitar syndrome.

CENTRAL MESSAGE

This review deals with the natural and surgical history of patients with scimitar syndrome, describing the clinical presentation and possible results based on the choice of treatment.

See Commentary on page 81.

ANATOMICAL AND PHYSIOLOGICAL SPECTRUM

The embryological origin of the syndrome is still unknown and may be related to a right/left disturbed signaling.[15] The typical scimitar vein (SV) usually provides drainage for the entire lung in 60% of cases. This vein may be stenotic in 20% of patients with a supra or infradiaphragmatic course, representing an important factor of poor prognosis.[15] Right lung hypoplasia (70% of cases) of varying degrees can be identified and is typically associated with dextrocardia (50% of cases) and systemic arterial supply (50% of cases) (Table 1). Atrial septal defect is present in 50% of the patients, but other complex congenital heart diseases (CHDs) may occur in up to 20% of patients. The remaining patients have isolated SS forms (approximately 40% of cases).[14]

The anomalous venous return inevitably leads to a left-to-right shunt, with an increase in the pulmonary-systemic flow (Qp/Qs) ratio and consequent right ventricular overload and dilation. Pulmonary arterial hypertension may occur in 30% of cases and is observed mainly in neonates/infants, in patients with CHD, and more rarely in isolated SS forms. This factor is widely recognized as one of the main determinants of poor prognosis.[8,9,14]

The diagnosis generally includes an echocardiographic evaluation in almost all patients, catheterization in more

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TABLE 1. Overall demographic, clinical, and instrumental data and outcomes according to the age of patients at diagnosis of a series of patients with a diagnosis of scimitar syndrome (n = 485)

		Age groups (at diagnosis)			
	Overall	Neonate/infants (0-1 y)	Children (≥1-10 y)	Adolescents/adults (≥10 y)	*P* value
Number of patients*	485 (100%)	n = 282 (58%)	n = 113 (23%)	n = 90 (19%)	
Sex, male*	180 (37%)	116 (41%)	43 (38%)	21 (23%)	.03
Heart position					.05
Dextrocardia*	240 (49%)	148 (52%)	61 (54%)	31 (34%)	
Mesocardia*	83 (17%)	48 (17%)	19 (17%)	16 (18%)	
Levocardia*	163 (33%)	86 (30%)	33 (29%)	43 (48%)	
Isolated forms*	186 (38%)	81 (29%)	57 (50%)	48 (53%)	.003
Associated CHD*	299 (62%)	199 (71%)	60 (53%)	40 (44%)	.003
Simple CHD*	198 (41%)	116 (41%)	49 (43%)	33 (37%)	
Complex CHD*	101 (21%)	83 (29%)	11 (9.7%)	7 (7.8%)	
Atrial septal defect*	243 (50%)	156 (55%)	56 (46%)	35 (39%)	
Right pulmonary hypoplasia*	346 (71%)	220 (78%)	76 (67%)	50 (55%)	.003
Degree of right pulmonary hypoplasia					.003
Mild*	204 (59%)	115 (52%)	50 (66%)	39 (78%)	
Moderate*	96 (28%)	65 (30%)	21 (28%)	10 (20%)	
Severe*	46 (13%)	40 (18%)	5 (6%)	1 (2%)	
Symptoms at diagnosis*	353 (73%)	220 (78%)	73 (65%)	60 (67%)	.047
Cardiac symptoms*	227 (47%)	168 (60%)	29 (27%)	30 (33%)	.006
Respiratory symptoms*	243 (50%)	140 (50%)	59 (52%)	44 (49%)	.99
Dilated RV at diagnosis (at 2D echo)*	332 (68%)	186 (66%)	84 (74%)	62 (69%)	.99
Cardiac catheterization*	276 (57%)	154 (55%)	75 (66%)	47 (52%)	.24
Qp/Qs†	2.1 (1.5-2.6)	2.1 (1.4-2.7)	1.8 (1.5-2.3)	2.1 (1.6-2.2)	.99
mPAP, mm Hg†	24 (18-34)	26 (20-40)	20.5 (17-30)	24 (18.5-28)	.006
Pulmonary hypertension*	157 (32%)	112 (40%)	28 (25%)	17 (19%)	.006
SAS to the right lung*	248 (51%)	182 (65%)	49 (43%)	17 (19%)	.006
Embolization of SAS*	177 (71%)	132 (47%)	33 (29%)	12 (13%)	.006
STPs*	279 (58%)	142 (51%)	78 (69%)	59 (66%)	.03
CMPs*	206 (42%)	140 (49%)	35 (31%)	31 (43%)	
Time to last follow-up, y†	7.2 (2.2-14.0)	6.4 (1.8-12.7)	9.9 (3.7-17.0)	7.5 (2.3-12)	.02
Overall mortality*	41 (8.7%)	37 (13%)	4 (3.5%)	–	.01
STPs hospital mortality*	29 (10%)	25 (17%)	4 (5%)	–	.08
STPs late mortality*	11 (3.9%)	10 (7%)	1 (1.3%)	–	.08
CMPs mortality*	13 (6.3%)	13 (9.3%)	–	–	.14
Symptoms at follow-up*	172/451 (37%)	109 (42%)	33 (31%)	30 (35%)	.24

CHD, Congenital heart disease; *RV*, right ventricle; *2D*, 2-dimensional; *Qp/Qs*, pulmonary-systemic flow ratio; *mPAP*, mean pulmonary artery pressure; *SAS*, systemic arterial supply; *STPs*, surgically treated patients; *CMPs*, clinically monitored patients. *Number of patients and percentage. †Median and interquartile range. Adapted with permission from Vida and colleagues.[14]

than 50% of them and, more rarely, computed tomography and magnetic resonance imaging (30% of cases). Catheterization also allows the occlusion of aberrant systemic arterial supply by embolization coils, if required by hemodynamic conditions. From an electrocardiographic point of view, 50% of cases show right ventricular hypertrophy, in the form of a right bundle branch block. Pulmonary scintigraphy may be performed and usually shows that the left lung is hyperperfused compared with the right one (76% vs 24%).

CLINICAL PRESENTATION AND MEDICAL TREATMENT

The clinical presentation of SS varies and is closely related to the associated CHDs. In about 60% of cases, the diagnosis is made in children/neonates younger than

TABLE 2. Demographics and surgical data of a series of patients with scimitar syndrome undergoing surgery (n = 279)

	Mean ± SEM or n
Demographics	
Male patients	109/279 (39%)
Age at surgery, mo	10.2 ± 0.9
Group of age at diagnosis	
<1 y	142/279 (51%)
>1 y; <10 y	78/279 (28%)
> 10 y	59/279 (21%)
Characteristics	
Symptoms at diagnosis	210/279 (76%)
Cardiac symptoms at diagnosis	147/279 (53%)
Respiratory symptoms at diagnosis	142/279 (51%)
Need for medications at diagnosis	48/279 (17%)
Pulmonary hypertension at diagnosis	105/279 (38%)
mPAP at diagnosis, mm Hg	29.2 ± 0.1
Associated CHDs	194/279 (69%)
Atrial septal defect	168/279 (60%)
Complex CHDs	63/279 (23%)
SAS to the right lung	130/279 (47%)
Coil embolization of SAS	93/279 (33%)
Degree of right pulmonary hypoplasia	
Mild	110/279 (39%)
Moderate	61/279 (22%)
Severe	18/279 (6%)
Operative data	
Type of procedure	
Corrective surgery	254/279 (91%)
Resective	25/279 (9%)
Type of corrective surgery	
Intracardiac baffle	176/254 (69%)
Direct scimitar vein reimplantation	78/254 (31%)
Type of resective surgery	
Lobectomy	18/25 (72%)
Pneumectomy	7/25 (28%)
Associated surgical procedure	101/279 (36%)
CBP time, min	127.2 ± 5.4
Aortic crossclamp time, min	62.5 ± 3.2
Circulatory arrest time, min	28.3 ± 6.4

SEM, Standard error of the mean; *mPAP*, mean pulmonary artery pressure; *CHD*, congenital heart disease; *SAS*, systemic arterial supply; *CBP*, cardiopulmonary bypass.

the first year of age. This form, previously known as ''infantile form''[9] is more frequently characterized by symptoms at diagnosis, such as congestive heart failure (60% of patients), failure to thrive, and tachypnea.[14] In addition, 50% of these patients have respiratory symptoms, such as difficulty breathing, recurrent upper respiratory tract infections, cyanosis, and pneumonia. This group is also frequently affected by the presence of collateral arteries that supply the right lung and by associated CHDs. For this reason, these patients are more likely to be diagnosed early in life and treated surgically.

In contrast, an ''adult form'' has been identified as a milder manifestation of the syndrome.[8] In our case series, we have divided this spectrum into 3 different subcategories based on age: neonates within 1 year, children from 1 to 10 years, and adolescents/adults older than 10 years. Fewer children were asymptomatic at diagnosis (65% of cases), with a very low percentage of cardiac symptoms (27% of cases) and pulmonary hypertension (25% of cases) (Table 1). Interestingly, we have confirmed that in more than 50% of adolescents/adults, SS appears as an isolated form, with a slight degree of right pulmonary hypoplasia, absence of symptoms in 33% of patients, or history of recurrent upper respiratory tract infections in 50% of them.[14]

Medical treatment in patients who are not directed at surgery (ie, clinically monitored patients [CMPs]) depends on the presence and type of symptoms at diagnosis. Cardiac symptoms are generally managed with diuretics and Na^+/K^+ ATPase inhibitors, whereas respiratory symptoms may require drugs such as beta-agonist and ipratropium bromide, antibiotics, and cortisonics. As mentioned previously, patients may present with an abnormal systemic supply to the right lung that can be treated with coil embolization, to reduce the Qp/Qs ratio.

SURGICAL MANAGEMENT

More than one half of patients (57%) are treated surgically (ie, surgically treated patients [STPs]), with an average age of about 10 years at surgery. Neonates patients are diagnosed earlier (51% of cases), particularly in the presence of cardiac symptoms and associated CHDs[14] (Tables 1 and 2). Pulmonary hypertension (defined as a the presence of a mean pulmonary artery pressure >25 mm Hg at rest or 50% above systemic level) is often present before surgery (38% of patients) and must always be addressed before surgery, either by medical treatment or by reducing the Qp/Qs ratio by coil embolization. Therefore, isolated forms of SS are generally treated in the case of a Qp/Qs ratio >1.5:1 or Qp/Qs ratio <1.5:1 with a clinically treated pulmonary hypertension. However, the right choice for this particular group of patients remains controversial.

Surgical treatment includes correction of abnormal venous drainage in most patients (>90%), but right lung resection may be required in more severe forms. In any case, nonetheless, the abnormal systemic supply to the right lung should be ligated and all associated CHDs should be repaired.

Surgical options for the redirection of the anomalous SV in the left atrium can consist of intracardiac baffle (69% of cases) or direct SV reimplantation (31% of cases).[14] There are several surgical corrective options for the pulmonary venous return re-routing,[16] which can be accomplished both via a right thoracotomy or sternotomy (Figure 1, Video 1). One of the first options described involved the creation of a pericardial tunnel

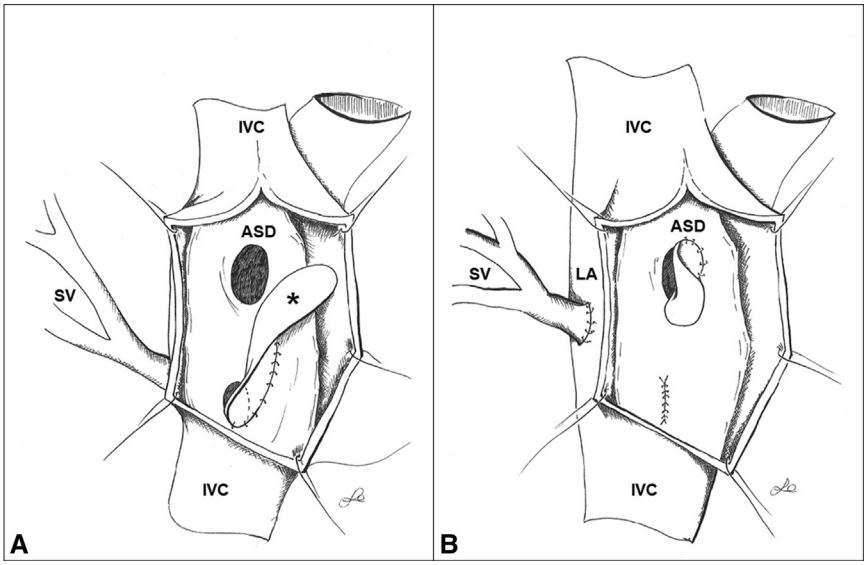


FIGURE 1. A, Intracardiac baffle technique. B, Reimplantation technique. The *asterisk* shows the intracardiac baffle. *SV*, Scimitar vein; *ASD*, atrial septal defect; *IVC*, inferior vena cava; *LA*, left atrium.

baffling flow from the orifice of the SV at the level of the IVC through the right atrium and then the connection of the baffle to the sides of the atrial septal defect[17] (Figure 1, *A*). Another technique includes the resection of the SV orifice with a cuff of the IVC and subsequent reimplantation in the left atrium and IVC patch enlargement[18] (Figure 1, *B*). Brown and colleagues[19] described a direct anastomosis of the SV, with full dissection of the SV, incision of the diaphragm, and reimplantation through a window excised posterior to the phrenic nerve. Recently, new techniques have been proposed, such that by Lugones and García,[20] in which the pulmonary venous return is widely connected to the left atrium through a tunnel constructed with the in situ pericardium.

Nowadays, the role of a right pulmonary lobectomy or more rarely a right pulmonary pneumectomy has been limited to the presence of severe right pulmonary hypoplasia, recurrent upper respiratory tract infections unresponsive to medical treatment, diffuse bronchiectasia, persistent hemoptysis, or intra-atrial baffles thrombosis after corrective surgery.

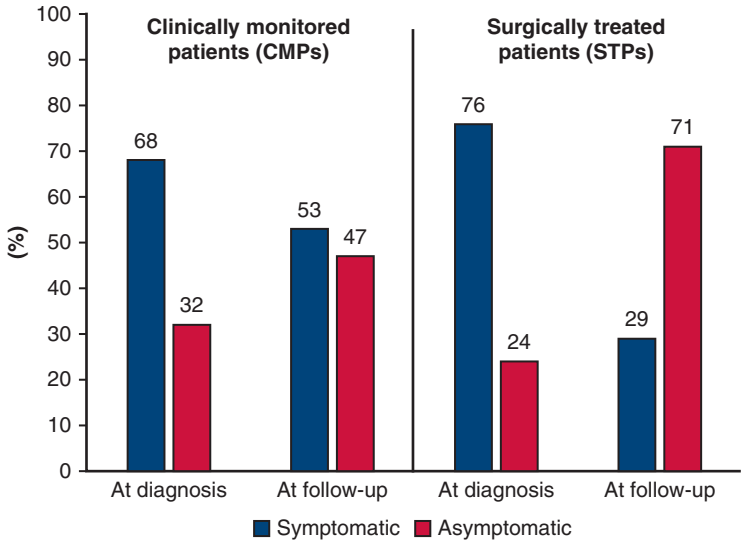


FIGURE 2. Bar plots showing the difference in the presence of symptoms at the time of diagnosis and at the time of follow-up between clinically monitored patients (*CMPs*) and surgically treated patients (*STPs*). The associations among clinical variables and outcome measures are based on a logistic regression model for binary outcomes (odds ratio, 0.16; confidence interval, 0.08-0.32).

TABLE 3. Outcomes of a series of scimitar syndrome patients undergoing surgery (n = 279)

	Mean ± SEM or n
Early outcomes	
Postoperative complications	93/279 (33%)
Respiratory failure	36/93 (39%)
Congestive heart failure	21/93 (26%)
Sepsis	16/93 (17%)
Pericardial/pleural effusion	14/93 (15%)
Arrhythmia	14/93 (15%)
Pulmonary hypertensive crisis	14/93 (15%)
Bleeding	8/93 (9%)
Delayed chest closure	6/93 (6%)
Hospital mortality	17/279 (6%)
Hospital mortality in case of corrective surgery	8/254 (3%)
Hospital mortality in case of resective surgery	9/25 (36%)
Late outcomes	
Late mortality	11/279 (4%)
Overall mortality	28/279 (10%)
Congestive heart failure	16/28 (57%)
Pulmonary hypertension	6/28 (22%)
Respiratory failure	4/28 (14%)
Bleeding	2/28 (7%)
Symptoms at follow-up	80/279 (29%)
Cardiac symptoms at follow-up	49/279 (18%)
Respiratory symptoms at follow-up	31/279 (11%)
Need for medications at follow-up	52/279 (19%)
Scimitar vein anastomosis presenting	
Stenosis	57/254 (22%)
Occlusion	6/254 (2%)
Reoperation/recatheterization in case of corrective surgery	42/254 (17%)
Freedom from above, y	6.3 ± 0.4

SEM, Standard error of the mean.

Postoperative complications occur in 33% of the patients and usually include respiratory failure (39%) and congestive heart failure (26%). The hospital mortality rate is generally low (6%) and lower for patients undergoing corrective surgery (3%), rather than resective procedures (36%). Late mortality is around 4% at a mean follow-up of 7 years, with an overall mortality of 10% of the patients.

LATE SURGICAL OUTCOMES VERSUS NATURAL HISTORY

Overall survival probability at 30 years of age is 88% and is lower in patients with associated CHDs and pulmonary hypertension. In the STPs, most of the deaths are caused by congestive heart failure (57%), but also by pulmonary hypertension (22%) and respiratory failure (14%).

Most STPs are asymptomatic at the last clinical examination (71%) (Figure 2) and only 20% of patients still need medications (Table 3). In contrast, there is a slight increase in symptomatic patients in clinically monitored patients due to both for cardiac and respiratory symptoms. None of the asymptomatic patients at diagnosis develops symptoms after surgical treatment.[14]

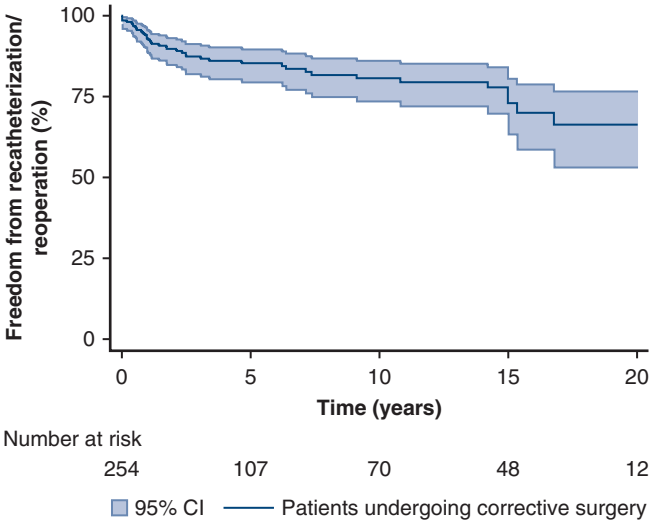


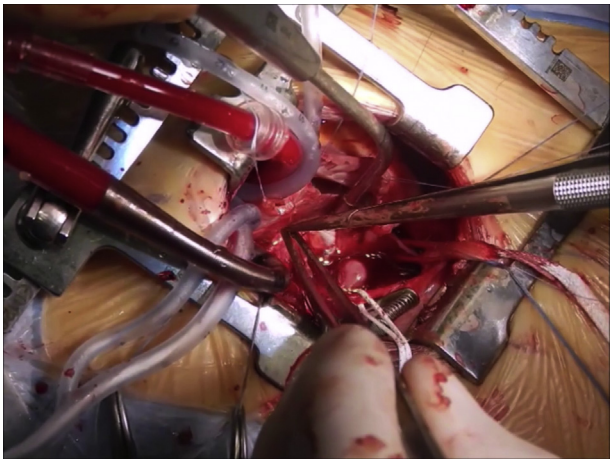
FIGURE 3. Kaplan–Meier curves showing the freedom from interventional cardiac catheterization or reoperation on "scimitar vein" anastomosis after corrective surgery (n = 254 patients). *95% CI*, 95% confidence interval.

Nevertheless, almost one quarter of the STPs present SV drainage disfunction after a corrective procedure. This included stenosis in 22% of cases or rarely total occlusion in 2% of patients. This diagnosis is not related to the type of corrective technique[14] used and is more frequent in neonates/infants (33%) than in either children (20%) or adolescents/adults (14%). In addition, we found that there is an inverse linear correlation between age at correction and stenosis/occlusion of the scimitar drainage (the younger the patient, the higher the incidence of stenosis/occlusion).[14] For this reason, an intracardiac baffle is usually recommended in younger patients whereas direct reimplantation is recommended in older ones. Of these patients with SV occlusion, 17% need an interventional cardiac catheterization or reoperation to address the failure, with average freedom of more than 6 years from the time of the first surgery (Figure 3, Table 3). In some patients with obstruction of the SV baffle, a pneumonectomy is required, and hemoptysis is often the first indication of this complication.

CONCLUSIONS

The clinical presentation of patients with SS varies, mainly depending on age at diagnosis and associated CHDs. Pulmonary hypertension and the presence of associated CHDs are negative prognostic factors in patient survival, regardless of patient history.

Surgical treatment usually involves redirection of the anomalous pulmonary venous drainage into the left atrium. This usually provides excellent results with regards to symptom relief and is superior in reducing the risk of



VIDEO 1. Video showing the surgical correction of a patient with scimitar syndrome by direct reimplantation in the left atrium via a right thoracotomy. Video available at: https://www.jtcvs.org/article/S2666-2507(20)30044-4/fulltext.

developing late symptoms with respect to clinically monitored or medically treated patients.

However, almost 25% of the STPs undergoing corrective surgery present SV drainage disfunction, with stenosis or occlusion at the level of the SV anastomosis. This happens more frequently in young children and requires interventional cardiac catheterization or reoperation to treat related morbidities.

Treatment decisions are still challenging, especially in patients with less-severe forms who are often asymptomatic. New data obtained from large multicenter studies have shown that surgical treatment should be considered in isolated forms only when the scimitar drainage causes significant pulmonary overload. Correction of associated CHDs and occlusion of systemic arterial supply may be useful, thus postponing a full correction to an older age. Confirmatory prospective randomized clinical trials are necessary to reach more definitive conclusions.

Conflict of Interest Statement

Authors have nothing to disclose with regard to commercial support.

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