Scimitar Syndrome. A European Congenital Heart Surgeons Association (ECHSA) Multicentric Study

Vladimiro L. Vida, Massimo A. Padalino, Giovanna Boccuzzo, Erjon Tarja, Hakan Berggren, Thierry Carrel, Sertaç Çiçek, Giancarlo Crupi, Duccio Di Carlo, Roberto Di Donato, José Fragata, Mark Hazekamp, Viktor Hraska, Bohdan Maruszewski, Dominique Metras, Marco Pozzi, Rene Pretre, Jean Rubay, Heikki Sairanen, George Sarris, Christian Schreiber, Bart Meyns, Tomas Tlaskal, Andreas Urban, Gaetano Thiene and Giovanni Stellin

Circulation. published online September 7, 2010;
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

Print ISSN: 0009-7322. Online ISSN: 1524-4539

Scimitar Syndrome

A European Congenital Heart Surgeons Association (ECHSA) Multicentric Study

Vladimiro L. Vida, MD, PhD; Massimo A. Padalino, MD, PhD; Giovanna Boccuzzo, MPH; Erjon Tarja, MD; Hakan Berggren, MD; Thierry Carrel, MD; Sertaç Çiçek, MD; Giancarlo Crupi, MD; Duccio Di Carlo, MD; Roberto Di Donato, MD; José Fragata, MD; Mark Hazekamp, MD; Viktor Hraska, MD; Bohdan Maruszewski, MD; Dominique Metras, MD; Marco Pozzi, MD; Rene Pretre, MD; Jean Rubay, MD; Heikki Sairanen, MD; George Sarris, MD, PhD; Christian Schreiber, MD; Bart Meyns, MD; Tomas Tlaskal, MD; Andreas Urban, MD; Gaetano Thiene, MD; Giovanni Stellin, MD

Background—Scimitar syndrome is a rare congenital heart disease. To evaluate the surgical results, we embarked on the European Congenital Heart Surgeons Association (ECHSA) multicentric study.

Methods and Results—From January 1997 to December 2007, we collected data on 68 patients who underwent surgery for scimitar syndrome. Primary outcomes included hospital mortality and the efficacy of repair at follow-up. Median age at surgery was 1.4 years (interquartile range, 0.46 to 7.92 years). Forty-four patients (64%) presented with symptoms. Surgical repair included intraatrial baffle in 38 patients (56%; group 1) and reimplantation of the scimitar vein onto the left atrium in 21 patients (31%; group 2). Eight patients underwent right pneumectomy, and 1 had a right lower lobe lobectomy (group 3). Four patients died in hospital (5.9%; 1 patient in group 1, 2.6%; 3 patients in group 3, 33%). Median follow-up time was 4.5 years. There were 2 late deaths (3.1%) resulting from severe pulmonary arterial hypertension. Freedom from scimitar drainage stenosis at 13 years was 83.8% in group 1 and 85.8% in group 2. Four patients in group 1 were reoperated, and 3 patients (2 in group 1 [6%] and 1 in group 2 [4.8%]) required balloon dilation/stenting for scimitar drainage stenosis.

Conclusions—The surgical treatment of this rare syndrome is safe and effective. The majority of patients were asymptomatic at the follow-up control. There were a relatively high incidence of residual scimitar drainage stenosis that is similar between the 2 reported corrective surgical techniques used. **(*Circulation*. 2010;122:1159-1166.)**

Key Words: congenital heart disease ■ multicenter study ■ scimitar syndrome

Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies that has been reported in 3% to 6% of patients with partial anomalous venous connection. In these patients, some of the pulmonary veins drain into the upper part of the inferior vena cava,[1–6] either above or below the diaphragm,[7] and usually drain the lower and sometimes the middle lobe of the right lung (79%) or the whole right lung (21%).[1] The right lung is frequently hypoplastic and receives its blood supply from the systemic arteries, mainly the thoracic or abdominal aorta, and this supply is usually to the lower lobes.[8]

Clinical Perspective on p 1166

About 70% of these patients with scimitar syndrome have an associated atrial septal defect.[9] The syndrome also has

Received November 25, 2009; accepted June 28, 2010.

From the Departments of Pediatric and Congenital Cardiac Surgery (V.L.V., M.A.P., E.T., G.T., G. Stellin), Statistical Science (G.B.), and Medical Diagnostic Sciences and Special Therapies (G.T.), University of Padua, Padua Italy; Children's Heart Center, Queen Silvia Children's Hospital, Goteborg, Sweden (H.B.); Clinic for Cardiovascular Surgery, Swiss Cardiovascular Center, University Hospital, Bern, Switzerland (T.C.); Anadolu Medical Center, Istanbul, Turkey (S.C.); Department of Cardiac Surgery, Ospedali Riuniti, Bergamo, Italy (G.C.); Pediatric Cardiac Surgery Unit, Ospedale Pediatrico Bambino Gesù, Rome, Italy (D.D.C., R.D.D.); Hospital de Santa Marta, Seviço de Cirurgia Cardiotorácica Unidade Cardiáca Pediátrica, Lisbon, Portugal (J.F.); Department Cardiothoracic Surgery D6-S, Leiden University Medical Center, Leiden, the Netherlands (M.H.); German Pediatric Heart Center, Asklepios Clinic Sankt, Augustin, Germany (V.H., A.U.); The Children's Memorial Health Institute, Department of Cardiothoracic Surgery, Warsaw, Poland (B. Maruszewski); Department of Cardiothoracic Surgery Hopital de la Timone, Marseille, France (D.M.); Department of Pediatric and Congenital Cardiac Surgery and Cardiology, Ospedali Riuniti, Ancona, Italy (M.P.); Pediatric and Congenital Cardiac Surgery, University Children's Hospital, Zurich, Switzerland (R.P.); Service de Chirurgie Cardiovasculaire et Thoracique Université Catholique de Louvain, Bruxelles, Belgium (J.R.); Hospital for Children and Adolescents University of Helsinki, Helsinki, Finland (H.S.); Congenital Heart Surgery, Mitera Pediatric and Hygea Hospital, Athens, Greece (G. Sarris); German Heart Center, Munich Clinic of Cardiovascular Surgery, Munich, Germany (C.S.); Department of Cardiac Surgery, Leuven, Belgium (B. Meyns); and Department of Pediatric Cardiac Surgery, University Hospital, Prague, Czech Republic (T.T.).

Correspondence to Giovanni Stellin, MD, Pediatric and Congenital Cardiac Surgery Unit, University of Padua, Via Giustiniani, 2-35100 Padua, Italy. E-mail giovanni.stellin@unipd.it

Circulation **is available at http://circ.ahajournals.org** **DOI: 10.1161/CIRCULATIONAHA.109.926204**

been described less commonly in association with other cardiac malformations like tetralogy of Fallot,[10,11] ventricular septal defect,[7–9,12–15] coarctation of the aorta,[3,14,16] hypoplastic left heart syndrome,[5] total anomalous pulmonary venous connection,[16] patent ductus arteriosus,[3,10,13–18] cor triatriatum,[19] bicuspid aortic valve,[10,15] and subaortic stenosis.[16] Overall, 19% to 31% of patients with scimitar syndrome have associated cardiac anomalies.[1,10,11,15,20]

This syndrome can present early in the neonatal period or later in life with a wide clinical spectrum.[4,14,16,21] Dupuis et al[19] divided scimitar syndrome into 3 main forms: an infantile form with symptoms and pulmonary hypertension, an "older" adult form distinguished by being asymptomatic in infancy, and a form with associated congenital cardiac anomalies.

Surgical correction is usually accomplished in symptomatic patients or in patients with an increased pulmonary blood flow and signs of right heart chamber dilation.[4,13] Several different surgical approaches have been described over the years and are currently used, depending on the anatomic and pathological features of each case and according to the surgeon's preference.[15,19–28]

The objective of this study was to analyze the surgical results and follow-up outcome of patients who underwent surgery for scimitar syndrome within a European Congenital Heart Surgeons Association (ECHSA) multicentric data collection in a relatively recent time period. To the best of our knowledge, this is the widest surgical experience collected so far in the medical literature.

Methods

A review of the medical records and computerized hospital data was approved by the Clinical Investigation Committee of the University Hospital of Padua, and the procedures followed were in accordance to the institutional guidelines for retrospective record review and protection of patient confidentiality. Individual consent was not obtained by patients enrolled in this study. Patients were not identified, and the chairperson of the Ethics Committee of each institution consented to send data from that institution for publication.

The data reviewed were relative to the hospital course and to the follow-up clinical controls of patients with scimitar syndrome who required surgical treatment between January 1997 and December 2007. Nineteen of the 42 cardiothoracic centers within the ECHSA contributed to the data collection.

Variables analyzed included demographic and preoperative clinical data, different operative techniques (group 1, patients who had an intraatrial baffle repair; group 2, patients who had a reimplantation of the scimitar vein on the left atrium; and group 3, patients who underwent right pneumectomy or right lung lobectomy).

Primary outcomes included hospital mortality and morbidity rates, late mortality, and scimitar drainage patency at the last follow-up control. The presence of pulmonary arterial hypertension was defined as a mean pulmonary artery pressure >25 mm Hg at rest or >30 mm Hg with exercise.[29]

Quantitative variables were summarized as median and interquartile range (IQR), equal to the difference between the third (75% of the distribution) and first (25% of the distribution) quartiles. Because quantitative variables are not normally distributed, comparisons among groups were carried out by the Kruskal-Wallis test.[30] When 2 groups were compared, the Wilcoxon signed-rank test was used. The difference between proportions was assessed by the extension of the Fisher exact test proposed by Freeman and Halton[31] for comparisons among >2 proportions. Data were analyzed with SAS software, release 9.1.3 (SAS Stat 9.1, SAS Institute Inc, Cary, NC), and values of $P<0.05$ were considered significant.

Results

Sixty-eight consecutive patients who underwent surgical treatment for scimitar syndrome within 19 ECHSA centers were included in this study. Each center entered between 1 and 9 patients (median, 3 patients per center) in this study. There were 43 female patients (63%) and 25 male patients (37%). Preoperative diagnosis was established by echocardiography in all patients. Additional instrumental examinations included cardiac catheterization in 57 patients (84%) and MRI in 5 patients (7.3%). Forty-four patients (64%) presented with symptoms that included recurrent upper respiratory tract infections (n=27 patients), congestive heart failure (n=24), recurrent pneumonia (n=9 patients), and cyanosis (n=4 patients). The presence of congestive heart failure was significantly higher in patients <3 months of age compared with older patients ($P=0.02$). In a 46-day-old male patient scheduled for ventricular septal defect closure, an association of an anomalous right pulmonary venous drainage with the inferior vena cava was discovered accidentally at the time of repair.

Associated cardiac anomalies were present in 51 patients (75%). The most common included atrial septal defect secundum type (atrial septal defect II) in 44 patients (65%), ventricular septal defect in 11 (16%), and patency of the ductus arteriosus in 6 (9%). Nineteen of 68 patients (28%) presented with associated congenital heart disease, excluding patency of the ductus arteriosus and atrial septal defects. Right lung hypoplasia was described in 35 patients (52%).

Fifty-seven patients (84%) who underwent preoperative cardiac catheterization showed a median systolic pulmonary artery pressure of 30 mm Hg (IQR, 24 to 35 mm Hg). Thirteen patients (22%) presented with pulmonary arterial hypertension, which was reported more frequently in patients with other associated congenital heart defect. Median ratio of pulmonary blood flow to systemic blood flow was 2:1 (IQR, 1.8:1 to 3:1). Systemic arterial supply to the right lung was demonstrated in 20 patients (29%); 6 were treated by coil embolization of aortopulmonary collaterals at the time of preoperative study. All 6 patients presented with congestive heart failure ($P=0.001$). Pulmonary sequestration was reported in 6 patients (8.8%) (Table 1).

Surgical repair included intraatrial baffle repair in 38 patients (56%; group 1) and reimplantation of the scimitar vein on the left atrium in 21 patients (31%; group 2). Six patients who underwent surgical repair (3 in group 1 and 3 in group 2) underwent a simultaneous ligation of collateral arteries supplying the right lung. The remaining 9 patients underwent a right pneumectomy (8 patients) or a right lower lobe resection (1 patient; group 3). One patient in group 3 underwent a previous atrial septal defect closure followed by a right lung pneumectomy during the same hospitalization.

Median age at surgery was 1.4 years (IQR, 0.46 to 7.92 years). Thirty-one patients (45%) were <12 months of age, and 54 of 68 patients (79%) underwent surgery within the first decade of life. Surgical approach was through a midline sternotomy in 52 patients (76%) and a right thoracotomy in 16 patients (24%; 3 patients in group 1, 6 patients in group 2, and 7 patients in group 3). Patients in group 3 were significantly younger than patients in groups 1 and 2 ($P=0.05$;

Table 1. Patients' Characteristics According to Degree of Right Lung Involvement in the Scimitar Drainage

	Right Lower Lobe (n=21)	Right Middle and Lower Lobe (n=9)	Entire Right Lung (n=38)	*P*
Median age at surgery (IQR), y	5.64 (0.78–10.50)	3.19 (0.58–13.17)	0.72 (0.37–5.79)	0.09*
Preoperative cardiac catheterization, n (%)	19 (90.5)	6 (66.7)	31 (81.6)	0.26
Presence of arterial collateral supply requiring embolization, n (%)	0	0	6 (15.8)	0.12
Presence of extralobar sequestration, n (%)	2 (9.5)	0	4 (10.5)	0.85
Pulmonary arterial hypertension, n (%)	2 (9.5)	2 (22.2)	8 (21.0)	0.59
Preoperative CHF, n (%)	3 (14.3)	1 (11.1)	20 (52.6)	0.003
Right lung pneumectomy/right lower lobe lobectomy, n (%)	0	1 (11.1)	8 (21.0)	0.051
Hospital mortality, n (%)	0	1 (11.1)	3 (7.9)	0.44
Late mortality, n (%)	0	1 (12.9)	1 (2.9)	0.34

CHF indicates congestive heart failure. Percentages are relative to the total in each group. The level of significance is referred to the overall comparison. It is based on Kruskal-Wallis test for quantitative variables and on an extension of the Fisher exact test to compare >2 groups by Freeman and Halton[31] for dichotomous variables.

*In the comparison of the first 2 groups considered as a the whole and the third one, the significance with the Fisher exact test is $P=0.03$.

Tables 2 and 3). Associated surgical maneuvers were reported in 51 patients (75%) and are listed in Table 4.

Median time in the intensive care unit and hospitalization time are reported in Tables 1 and 2. Postoperative complications were reported in 35 patients (51%; Table 5), being more frequent in group 3 (7 of 9, 78%; $P=0.02$) compared with group 2 (14 of 21, 67%) and group 1 (14 of 38, 37%) and essentially related to the presence of preoperative congestive heart failure (odds ratio=13, $P=0.004$; Tables 1 and 2). Patients who had postoperative complications were younger (median age, 0.59 years; IQR, 0.33 to 2.37 years) compared with uncomplicated patients (median age at surgery, 5.6 years; IQR, 0.9 to 12 years; $P=0.003$, Wilcoxon signed-rank test).

Patients in group 2 showed a significantly higher incidence of postoperative arrhythmias ($P=0.002$, extended Fisher exact test) and phrenic nerve injury ($P=0.04$, extended Fisher exact test). Patients in group 3 had a significantly higher incidence of prolonged intubation (>7 days; $P=0.01$, extended Fisher exact test) and postoperative pneumonia ($P=0.001$, extended Fisher exact test).

Four patients (5.9%) died in hospital: 1 of severe brain injury 56 days after an initial intraatrial baffle repair (group 1, 2.6%) and 3 in group 3 (33%; $P=0.008$) 15, 35, and 157 days after right pneumectomy for neurological problems, pulmonary hypertensive crises, and sepsis, respectively. Hospital mortality was significantly higher in patients with pulmonary

Table 2. Patients' Characteristics and Outcomes According to Surgical Techniques

	Group 1* (n=38, 56%)	Group 2† (n=21, 31%)	Group 3‡ (n=9, 13%)	*P*
Median age at surgery (IQR), y	2.26 (0.57–8.53)	1.16 (0.45–6.48)	0.46 (0.26–0.67)	0.46
Males, n (%)	15 (39.5)	8 (38.1)	2 (22.2)	0.72
Median body weight at surgery (IQR), kg	13 (5–39)	7 (4–18)	3 (3–6)	0.01
Median CPB time (IQR), min	123 (100–142)	120 (104–172)	107 (82–154)	0.68
Circulatory arrest time, n (%)	20 (52.6)	13 (61.9)	3 (33.3)	0.40
Median circulatory arrest time (IQR), min	48 (35–60)	67 (60–85)	14 (7–47)	0.0041
Hospital mortality, n (%)	1 (2.6)	0	3 (33.0)	0.0062
Postoperative complications, n (%)	14 (36.8)	14 (66.7)	7 (77.8)	0.0228
Median ICU stay (IQR), d	3 (2–7)	5 (2–8)	17 (6–35)	0.018
Median hospitalization (IQR), d	12.5 (7–18)	20 (10–27)	20 (15–35)	0.04
Late mortality, n (%)	1 (2.7)	1 (4.8)	0	1
Stenosis of the SV, n (%)	6 (16.2)	3 (14.3)	0	1
Reoperations for SV stenosis, n (%)	4 (10.8)	0	0	0.28
Hemodynamic procedures for SV stenosis, n (%)	2 (5.6)	1 (4.8)	0	1

CPB indicates cardiopulmonary bypass; SV, scimitar vein. Percentages are relative to the total in each group. The level of significance is referred to the overall comparison. It is based on Kruskal-Wallis test for quantitative variables and on an extension of the Fisher exact test to compare >2 groups by Freeman and Halton[31] for dichotomous variables. Pairwise comparisons between procedures are reported in Table 3.

*Group 1, intraatrial baffle to the left atrium; †group 2, direct reimplantation of the scimitar vein to the left atrium; ‡group 3, pneumonectomy/right lobe lobectomy.

Table 3. Significance of Pairwise Comparison Between Surgical Techniques

	Group 1* vs 2†	Group 1* vs 3‡	Group 2† vs 3‡
Median age at surgery	0.58	0.05	0.11§
Males	1	0.45	0.67
Median body weight at surgery	0.18	0.005	0.04
Median CPB time	0.55	0.74	0.37
Circulatory arrest time	0.59	0.46	0.24
Median circulatory arrest time	0.008	0.07	0.02
Hospital mortality	1	0.02	0.02
Postoperative complications	0.03	0.06	0.68
Median ICU stay	0.25	0.009	0.03
Median hospitalization	0.05	0.04	0.51
Late mortality	1	1	1
Stenosis of the SV	1	. . .	. . .
Reoperations for SV stenosis	0.29	. . .	. . .
Hemodynamic procedures for SV stenosis	1	. . .	. . .

CPB indicates cardiopulmonary bypass; ICU, intensive care unit; and SV, scimitar vein.

*Group 1, intraatrial baffle to the left atrium; †group 2, direct reimplantation of the scimitar vein to the left atrium; ‡group 3, pneumonectomy/right lobe lobectomy. The level of significance is referred to the Wilcoxon rank test for quantitative variables and Fisher exact test for dichotomous variables.

§In the comparison between the first 2 groups considered as a whole and the third one, the significance with the Fisher exact test is $P=0.05$.

arterial hypertension ($P=0.03$; Figure 1). All patients who died in hospital were found to have preoperative congestive heart failure and severe respiratory symptoms.

All patients who underwent a primary right lung pneumectomy had severe right lung hypoplasia, and their scimitar vein drained the entire right lung (Table 1). Five of 9 patients had preoperative congestive heart failure (56%), and 5 patients (56%) had severe respiratory symptoms. In addition, the majority of patients presented with a severe degree of pulmonary arterial hypertension at the catheterization study (5 of 9, 56%; $P=0.003$).

Table 4. Associated Surgical Procedures in 51 Patients (75%)

Procedure	n
Atrial septal defect closure	44
Ventricular septal defect closure	12
Patent ductus arteriosus ligation	6
Ligation of collateral arteries	6
Plasty of pulmonary artery branches	3
Correction of anomalous left PVC	2
Tricuspid valve plasty	2
Mitral valve plasty	1
Total cavopulmonary connection	1
ALCAPA reimplantation	1
Cox-Maze procedure	1
Pulmonary valve commissuroplasty	1
Pulmonary vein plasty	1

PVC indicates pulmonary venous connection; ALCAPA, anomalous origin of the coronary artery from the pulmonary artery.

Median follow-up time was 4.5 years (IQR, 1.6 to 8.3 years), which was completed in 63 of the 64 survivors (98.5%). One patient who underwent right pneumectomy was lost to follow-up.

There were 2 late deaths (3.1%), both resulting from right ventricular failure caused by severe pulmonary arterial hypertension (1 in group 1, 2%; 1 in group 2, 4%; $P=0.58$) 8 years and 126 days after surgery, respectively (Figure 1). A lung biopsy obtained in the patient in group 2 at the time of scimitar vein reimplantation, ventricular septal defect closure, and patency of the ductus arteriosus ligation showed Heath-Edwards class III pulmonary vascular obstructive disease.

At the last clinical follow-up control, 52 of 61 patients (85.2%) were in New York Heart Association class I. Among the 9 remaining patients, all in New York Heart Association class II, symptoms were consistent with persistent dyspnea and recurrent respiratory infection.

Outcomes according to the Dupuis classification[19] of scimitar syndrome are listed in Table 6. After corrective procedures, 9 patients (15.5%) showed some degree of stenosis of the scimitar drainage anastomosis at the last follow-up control, which was confirmed by angiography in all patients (6 of 37 in group 1, 16.2%; 3 of 21 in group 2, 14.2%; Figure 2). The presence of a scimitar drainage stenosis was correlated with younger age at repair (<3 months of age) only in patients in group 2 ($P=0.07$).

Four patients (7%), all in group 1, underwent reoperation (consisting of repositioning of the intraatrial baffle) and survived; symptoms (dyspnea and fatigue) were still present in 1 patient. Three patients (5.3%) underwent balloon dilation at the level of the stenotic connection (plus stent implantation in one of them) 8, 12, and 15 months after correction, respectively (2 in group 1, 6%; 1 in group 2, 4.8%). One patient (group 1) is still symptomatic for recurrent respiratory tract infections.

Three patients with instrumental diagnosis of scimitar drainage stenosis (1 patient in group 1 and 2 patients in group 2) are completely asymptomatic. So far, they have not been treated.

At multivariate analysis (logistic regression), no variables were significantly associated with outcomes.

Discussion

Scimitar syndrome is a rare congenital heart disease characterized by a wide spectrum of symptoms depending mainly on the presence of other associated congenital heart malformations, the amount of blood draining into the inferior vena cava, the presence of scimitar vein obstruction, the degree of the arterial supply to the right lung or its lower lobe, and the presence of the bronchial abnormalities responsible for retained secretions, lobar infections, and hemoptisis.[2,4,5,7,10,15,20]

Because of the wide range of symptoms at clinical onset, the diagnosis may be difficult, especially in children and young

Table 5. Postoperative Complications According to Surgical Procedure (66 Complications in 35 Patients)

	Group 1* (n=14 of 38 Patients, 37%), n	Group 2† (n=14 of 21 Patients, 67%), n	Group 3‡ (n=7 of 9 Patients, 78%), n	Total, n
Prolonged intubation (>7 d)	3	1	4	8
Pulmonary hypertensive crises	3	2	2	7
Low output syndrome	2	4	1	7
Arrhythmia	1	5	0	6
Phrenic nerve injury	1	4	0	5
Sternum left open	3	1	0	4
Pleural effusion requiring drainage	3	2	0	5
Reoperation during same admission	2	1	2	5
Pneumonia	0	0	3	3
Septicemia	2	1	1	4
AV block rep. temporary pace maker	2	1	0	3
Pneumothorax	2	0	0	2
Pericardial effusion requiring drainage	1	1	0	2
Neurological complications	1	0	1	2§
Tracheostomy	0	0	1	1
Cardiac arrest	0	0	1	1
Chylothorax	0	1	0	1

AV indicates atrioventricular.

*Group 1, intraatrial baffle to the left atrium; †group 2, direct reimplantation of the scimitar vein to the left atrium; ‡group 3, pneumonectomy/right lobe lobectomy.

§Both had deep hypothermic circulatory arrest.

adults with concomitant congenital heart lesions.[2–5,32,33] Pulmonary artery hypertension often occurs and is seen mostly in infants[12,14,16–18,20] with associated congenital heart malformations or with an anomalous large systemic arterial supply.[11,34–37] However, it has also been reported in adult patients.[38] A less common cause of pulmonary arterial hypertension is the presence of pulmonary vein stenosis.[16,17,20]

If the presence of a large systemic anomalous arterial supply to the right lung with pulmonary overcirculation is detected preoperatively, it should be abolished by occluding the systemic arteries that supply the affected lobe(s), thereby reducing the blood flow and the amount of shunting.[15,16,20,32–34,37–40] This has been advocated as the best and simplest treatment, especially in infants with congestive heart failure, often improving the clinical symptoms and decreasing the pulmonary arterial pressure.[1,14,17–19]

The surgical repair of scimitar syndrome consists of redirecting the pulmonary venous drainage into the left atrium,[14,15] either baffling the anomalous drainage into the left atrium via a tunnel or transecting the "scimitar drainage" near its entrance into the inferior vena cava and then reimplanting it directly into the left atrium.[24] Both baffle repair and direct reimplantation of the scimitar vein have been reported for many years and are currently used according to surgeon preference and to the anatomic and pathological features of each case.[22–28] Still, there is no consensus as to which is the best surgical treatment option.

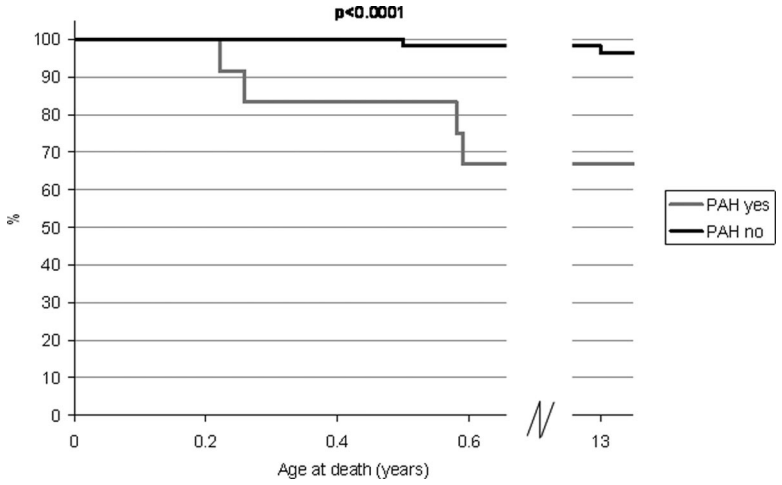

Figure 1. Kaplan-Meier estimate of overall survival according to the presence of pulmonary arterial hypertension (PAH).

Table 6. Outcomes According to Different Forms of Scimitar Syndrome

	Infantile Form* (n=38, 56%)	Adult Form† (n=11, 16%)	With Associated CHD‡ (n=19, 28%)	*P*
Median age at surgery (IQR), y	1.5 (0.47–5.6)	22 (17.5–52.4)	0.27 (0.16–0.76)	<0.0001
Pulmonary hypertension, n (%)	9 (23.7)	0	3 (15.8)	0.18
Preoperative CHF, n (%)	15 (39)	2 (18)	7 (37)	0.4
Postoperative complications, n (%)	19 (50)	3 (27)	13 (68)	0.1
Hospital mortality, n (%)	3 (7.9)	0	1 (5.3)	0.6
Late mortality, n (%)	1 (2.9)	0	1 (5.6)	0.40

CHD indicates congenital heart defects; CHF, congestive heart failure. Percentages are relative to the total in each group. The level of significance is referred to the overall comparison. It is based on the Kruskal-Wallis test for quantitative variables and on an extension of the Fisher exact test to compare >2 groups by Freeman and Halton[31] for dichotomous variables.

*An infantile form with symptoms and pulmonary hypertension.

†An "older" adult form distinguished by being asymptomatic in infancy.

‡A form with associated congenital heart defects that are the main determinant of this group (see Reference 19).

Although repair of the anomalous venous return and ligation of collaterals is generally recommended, Huddleston et al[14] reported that in symptomatic patients with repeated pulmonary infections and hemorrhage, a right lung lobectomy or pneumonectomy (either as primary therapy or if postoperative stenosis is present after failed repair) is an option leading to similar results.

In general, right lung pneumectomy is well tolerated in children,[41,42] particularly in patients in whom the affected lung is hypoplastic. Lobectomy can be done only if the anomalous vein drains only that lobe, and care needs to be taken to ensure that the remaining lung function will remain adequate.

In our multicentric ECHSA study, we report on a relatively large experience with the aim of analyzing the different surgical strategies, surgical outcomes, risks, and midterm results at follow-up. Our data indicate that the scimitar syndrome is not a simple and benign congenital heart disease and that it is often associated with other cardiac anomalies that play an important role in the long-term results.

The majority of our patients presented with preoperative symptoms that represent one of the main indications for surgical repair, particularly when pulmonary arterial hypertension is present. Almost all patients with severe associated anomalies presented within the first decade of life with symptoms that are difficult to ascribe to the anomalous scimitar vein drainage, the presence of associated anomalies, or both.

Patients who underwent right pneumectomy were significantly younger compared with patients who had corrective surgery, also showing a more severe degree of preoperative respiratory symptoms and congestive heart failure. In addition, the majority of patients presented with a severe degree of pulmonary arterial hypertension at the catheterization study. These data were consistent with the higher incidence of postoperative mortality and morbidity, confirming the experience in previous studies.[1,15,41,42]

The analysis of our data demonstrated that corrective surgery can be done safely with a low mortality and morbidity rate, which are independent of the type of corrective

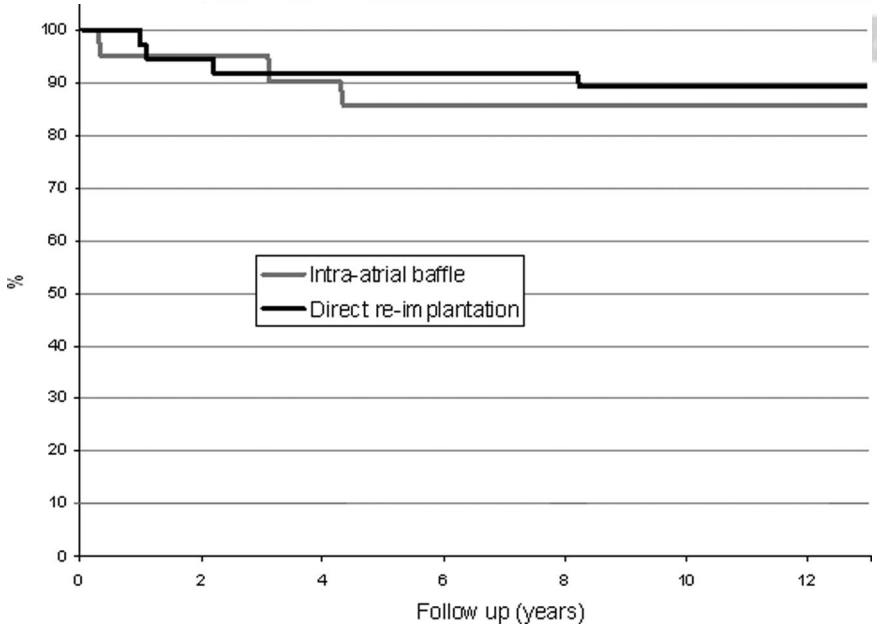

Figure 2. Kaplan-Meier estimate of freedom from scimitar drainage stenosis after surgical repair (according to surgical technique).

surgical technique used. Of note, the presence of pulmonary arterial hypertension is associated with a higher mortality rate. Also notable is the fact that the direct reimplantation technique is linked to a higher incidence of postoperative complications and a longer hospital stay.

The great majority of our patients showed good clinical conditions at the last clinical follow-up. The overall freedom from scimitar vein stenosis was 84.5% at 10 years and was similar between patients who had a reimplantation of the scimitar vein and patients who underwent an intracardiac baffle repair (85.8% versus 83.8%). It is of note that a residual stenosis at the scimitar vein level was correlated with younger age at repair (<3 months of age) only in patients in group 2.

We have found that the results of surgical treatment are linked to patient age. As a general rule, patients <1 year of age who require treatment are usually very ill, bearing a relatively high operative mortality and complication rate, whereas those who are older have a better outcome both immediately and in the long term. Previous reports have shown that delayed surgical treatment, especially in infants with associated congenital heart lesions, can lead to irreversible pulmonary vascular obstructive disease, which can jeopardize the surgical outcome of these patients.[2,5,11,13,17]

Our study has some limitations. Postoperative right lung assessment by pulmonary scintigraphy was performed in only a few patients in our cohort; therefore, it is impossible to draw any conclusions about the right lung anatomy and functional status and therefore patients' clinical conditions. We believe that such a test should be performed routinely in all patients after scimitar syndrome repair to achieve an appropriate long-term follow-up.

We are aware that this study design does not allow us to reach strong conclusions about the optimal procedure for this disease. However, we have collected a complete series of data on a large series of patients who were been treated surgically in the context of a rare congenital heart malformation with many anatomic variants and different surgical option strategies.

Conclusions

Our multicentric ECHSA study highlights the efficacy and safety of the surgical treatment of this rare syndrome. The majority of patients were asymptomatic at the latest clinical follow-up. However, we found a relatively high incidence of residual scimitar drainage stenosis requiring reoperation or hemodynamic reintervention, which is similar in the 2 reported surgical techniques.

Disclosures

None.

References

1. Alsoufi B, Cai S, Van Arsdell GS, Williams WG, Caldarone CA, Coles JG. Outcomes after surgical treatment of children with partial anomalous pulmonary venous connection. *Ann Thorac Surg*. 2007;84:2020–2026.
2. Vida VL, Speggiorin S, Padalino MA, Crupi G, Marcelletti C, Zannini L, Frigiola A, Varrica A, Di Carlo D, Di Donato R, Murzi B, Bernabei M, Boccuzzo G, Stellin G. The scimitar syndrome: an Italian multicenter study. *Ann Thorac Surg*. 2009;88:440–444.
3. Espinosa-Zavaleta N, Jativa-Chavez S, Munos-Castellanos L, Zamora-Gonzales C. Clinical and echocardiographic characteristics of scimitar syndrome. *Rev Esp Cardiol*. 2006;59:284–288.
4. Pikwer A, Gyllstedt E, Lillo-Gil R, Jönsson P, Gudbjartsson T. Pulmonary sequestration: a review of 8 cases treated with lobectomy. *Scand J Surg*. 2006;95:190–194.
5. Schramel FM, Westermann CJ, Knaepen PJ, van den Bosch JM. The scimitar syndrome: clinical spectrum and surgical treatment. *Eur Respir J*. 1995;8:196–201.
6. Snellen HA, van Ingen HC, Hoefsmit EC. Patterns of anomalous pulmonary venous drainage. *Circulation*. 1968;38:45–63.
7. Folger GM. The scimitar syndrome: anatomic, physiologic, developmental and therapeutic considerations. *Angiology*. 1976;27:373–407.
8. Clements BS, Warner JO. Pulmonary sequestration and related congenital bronchopulmonary-vascular malformations: nomenclature and classification based on anatomical and embryological considerations. *Thorax*. 1987;42:401–408.
9. Shibuya K, Smallhorn JE, McCrindle BW. Echocardiographic clues and accuracy in the diagnosis of scimitar syndrome. *J Am Soc Echocardiogr*. 1996;9:174–181.
10. Kiely B, Filler J, Stone S, Doyle EF. Syndrome of anomalous venous drainage of the right lung to the inferior vena cava: a review of 67 reported cases and three new case in children. *Am J Cardiol*. 1967;29:102–116.
11. Gikonyo DK, Tandon R, Lucas RV Jr, Edwards JE. Scimitar syndrome in neonates: report of four cases and review of the literature. *Pediatr Cardiol*. 1986;6:193–197.
12. Kamiyama M, Kamata S, Usui N. Scimitar syndrome treated with pneumonectomy: a case associated with bronchospastic attack. *Pediatr Surg Int*. 2004;20:65–66.
13. Canter CE, Martin TC, Spray TL, Weldon CS, Strauss AW. Scimitar syndrome in childhood. *Am J Cardiol*. 1986;58:652–654.
14. Huddleston CB, Exil V, Canter CE, Mendeloff EN. Scimitar syndrome presenting in infancy. *Ann Thorac Surg*. 1999;67:154–159.
15. Najm HK, Williams WG, Coles JG, Rebeyka IM, Freedom RM. Scimitar syndrome: twenty years' experience and results of repair. *J Thorac Cardiovasc Surg*. 1996;112:1161–1168.
16. Gao YA, Burrows PE, Benson LN, Rabinovitch M, Freedom RM. Scimitar syndrome in infancy. *J Am Coll Cardiol*. 1993;22:873–882.
17. Dupuis C, Charaf LA, Brevière GM, Abou P. "Infantile" form of the scimitar syndrome with pulmonary hypertension. *Am J Cardiol*. 1993;71:1326–1330.
18. Dupuis C, Rey C, Godart F, Vliers A, Gronnier P. Scimitar syndrome complicated by stenosis of the right pulmonary vein: apropos of 4 cases. *Arch Mal Coeur Vaiss*. 1994;87:607–613.
19. Dupuis C, Charaf LA, Brevière GM, Abou P, Rémy-Jardin M, Helmius G. The "adult" form of the scimitar syndrome. *Am J Cardiol*. 1992;70:502–507.
20. Uthaman B, Abushaban L, Al-Qbandi M, Rathinasamy J. The impact of interruption of anomalous systemic arterial supply on scimitar syndrome presenting during infancy. *Catheter Cardiovasc Interv*. 2008;71:671–678.
21. Tjang YS, Blanz U, Kirana S, Körfer R. Scimitar syndrome presenting in adults. *J Card Surg*. 2008;23:71–72.
22. Dupuis C, Charaf LA, Abou CP, Brevière GM. Surgical treatment of the scimitar syndrome in children, adolescents and adults: a cooperative study of 37 cases. *Arch Mal Coeur Vaiss*. 1993;86:541–547.
23. Casha AR, Sulaiman M, Cale AJ. Repair of adult Scimitar syndrome with an intra-atrial conduit. *Interact Cardiovasc Thorac Surg*. 2003;2:128–130.
24. Yilmaz M, Senkaya I, Cengiz M. Direct reimplantation technique via a right minithoracotomy for scimitar syndrome: a case report. *Heart Surg Forum*. 2006;9:843–845.
25. Mason DP, Mihaljevic T, Mazzone PJ, Murthy SC, Rice TW. Extrapleural pneumonectomy for scimitar syndrome. *J Thorac Cardiovasc Surg*. 2006;132:704–705.
26. Calhoun RF, Mee RB. A novel operative approach to scimitar syndrome. *Ann Thorac Surg*. 2003;76:301–303.
27. Rajaii-Khorasani A, Kahrom M, Mottaghi H, Kahrom H. Surgical management of scimitar syndrome: an alternative approach. *Anz J Surg*. 2008;78:419–420.
28. Muta H, Akagi T, Iemura M, Kato H. Coil occlusion of aortopulmonary collateral arteries in an infant with scimitar syndrome. *Jpn Circ J*. 1999;63:729–731.

29. Barst RJ, McGoon M, Torbicki A, Sitbon O, Krowka MJ, Olschewski H, Gaine S. Diagnosis and differential assessment of pulmonary arterial hypertension. *J Am Coll Cardiol*. 2004;16:40S–47S.
30. Kruskal WH, Wallis AW. Use of ranks in one-criterion variance analysis. *J Am Stat Assoc*. 1952;47:583–621.
31. Freeman GH, Halton JH. Note on an exact treatment of contingency, goodness of fit and other problems of significance. *Biometrika*. 1951;38: 141–149.
32. Nedelcu C, Carette MF, Parrot A, Hammoudi N, Marsault C, Khalil A. Hemoptysis complicating scimitar syndrome: from diagnosis to treatment. *Cardiovasc Intervent Radiol*. 2008;31:S96–S98.
33. Zagol B, Book S, Krasuski RA. Late "adult form" scimitar syndrome presenting with "infant form" complications. *J Invasive Cardiol*. 2006; 18:E82–E85.
34. Blaysat G, Kachaner J, Villain E, Sidi D, Pedroni E, Piéchaud JF, David N. Scimitar syndrome in infants: physiopathology and therapeutic implications in 12 cases. *Arch Fr Pediatr*. 1987;44:245–251.
35. Haworth Sg, Sauer U, Bühlmeyer K. Pulmonary hypertension in scimitar syndrome in infancy. *Br Heart J*. 1983;50:182–189.
36. Ishibashi N, Takahashi Y, Kikuchi T, Kobayashi N, Suzuki R, Mizutani S. Case report of scimitar syndrome in infancy: intracardiac repair after coil embolization of systemic arterial supply to sequestered lung. *Kyobu Geka*. 2001;54:875–879.
37. Tummers RF, Lam J, Nijveld A, Marcelletti C, Losekoot G. An infant with the scimitar syndrome and pulmonary hypertension: successful surgical intervention. *Eur Heart J*. 1987;8:194–197.
38. Nedelcu C, Carette MF, Parrot A, Hammoudi N, Marsault C, Khalil A. Hemoptysis complicating scimitar syndrome: from diagnosis to treatment. *Cardiovasc Intervent Radiol*. 2008;31:S96–S98.
39. Mas C, Goh TH, Wilkinson JL. New interventional therapeutic approach for dual drainage of the scimitar vein. *Catheter Cardiovasc Interv*. 2000; 51:192–195.
40. Hoffman JIE. The natural and unnatural history of congenital heart disease. In: *Scimitar Syndrome*. Hoboken, NJ: Wiley-Blackwell. 2009: 161–166.
41. Laros CD, Westermann CJ. Dilatation, compensatory growth, or both after pneumonectomy during childhood and adolescence: a thirty-year follow-up study. *J Thorac Cardiovasc Surg*. 1987;93:570–576.
42. Stiles QR, Meyer BW, Lindesmith GG, Jones JC. The effects of pneumonectomy in children. *J Thorac Cardiovasc Surg*. 1969;58: 394–400.

CLINICAL PERSPECTIVE

Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies that has been reported in 3% to 6% of patients with partial anomalous venous connection. To analyze the surgical results and follow-up outcome of patients who underwent surgery for scimitar syndrome, we embarked on an European Congenital Heart Surgeons Association (ECHSA) multicentric data collection in a relatively recent time period. Our data indicate that scimitar syndrome is not a simple and benign congenital heart disease and that it is often associated with other cardiac anomalies that play an important role in the long-term results. The majority of our patients presented with preoperative symptoms that represent one of the main indications for surgical repair, particularly when pulmonary arterial hypertension is present. We demonstrated that corrective surgery can be done safely with a low mortality and morbidity rate, which are independent of the type of corrective surgical technique used. Patients who underwent a right pneumectomy had severe right lung hypoplasia and showed a more severe degree of preoperative respiratory symptoms, congestive heart failure, and pulmonary arterial hypertension, which was also consistent with their higher incidence of postoperative mortality and morbidity. Of note also is the fact that the presence of pulmonary arterial hypertension is associated with a higher mortality rate. At the latest clinical control, the majority of patients were asymptomatic; however, we found a relatively high incidence of residual scimitar drainage stenosis (15.5%) requiring reoperation or hemodynamic reintervention, which is similar in the 2 reported surgical techniques.