

Clinical and Echocardiographic Characteristics of Scimitar Syndrome

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Seven patients with scimitar syndrome underwent retrospective clinical and echocardiographic examination. The findings were: respiratory infection with dyspnea on moderate exercise in 90%, scimitar sign in 4 (57%), dextrocardia in 5 (71%), and atrial septal defect in five (71%), one of whom had patent ductus arteriosus. Overall, 2 patients had patent ductus arteriosus: one also had aortic coarctation and the other, a bicuspid aortic valve. Dilation of the right cavities was found in 5 (71%) and blunt edge in 5 (71%). In 2 patients, anomalous drainage was into the right atrium; in another 2, into the inferior vena cava; and in 3, towards the junction of the right atrium and the inferior vena cava. In 3 patients, drainage was obstructed. Six patients with cardiac abnormalities proceeded to surgery. Scimitar syndrome is a rare entity. In the large majority of patients (86%), diagnosis and follow-up can be performed echocardiographically.

Key words: *Adult congenital heart disease. Scimitar syndrome. Echocardiography.*

Aspectos clínicos y ecocardiográficos del síndrome de la cimitarra

Se realizó un estudio retrospectivo de 7 pacientes adultos con síndrome de la cimitarra mediante examen clínico y ecocardiográfico. Los resultados fueron: infección respiratoria y disnea de medianos esfuerzos en el 90%, signo de la cimitarra en 4 (57%), dextrocardia en 5 (71%), comunicación interauricular en 5 (71%), uno de ellos con persistencia de conducto arterioso; 2 con persistencia de conducto arterioso, 1 de ellos con coartación de aorta y otro con aorta bivalva; dilatación de cavidades derechas en 5 (71%) y borde romo en 5 (71%). En 2 pacientes, el colector drenó la aurícula derecha, en 2 la vena cava inferior y en 3 la unión de la aurícula con la vena cava inferior; 3 tuvieron obstrucción del colector. Seis pacientes fueron intervenidos quirúrgicamente.

El síndrome de la cimitarra es raro; su diagnóstico y su seguimiento se puede realizar mediante ecocardiografía en un alto porcentaje de casos (86%).

Palabras clave: *Cardiopatía congénita del adulto. Síndrome de la cimitarra. Ecocardiografía.*

INTRODUCTION

Scimitar syndrome is characterized by an anomalous connection of the right pulmonary veins to the suprahepatic portion of the inferior vena cava or to the right atrium above the junction with this vein. Associated anomalies include dextroposition of the heart due to dextrorotation, hypoplasia of the right lung, hypoplasia of the right pulmonary artery, systemic anomalies of the right lung arterial irrigation (aortopulmonary collateral arteries that can cause a left-

to-right shunt), bronchopulmonary sequestration with agenesis of the upper right or middle bronchus, as well as other extracardiac and cardiac anomalies. Twenty-five percent of patients present cardiac malformations, with the most frequent being an atrial septal defect.¹⁻⁴

The presumptive diagnosis is made on radiographic findings. Echocardiography allows identification of the anomalous drainage of the right pulmonary veins toward the curved collector, the presence of obstructions, and the connection site in the suprahepatic portion of the inferior vena cava. The transthoracic technique has diagnostic limitations in up to 33% of cases; hence, it is important to perform a transesophageal study as well.^{5,6} The interest of this report lies in the fact that cases reported among the adult population are rare.

The aim of this study is to report the clinical and echocardiographic findings in a series of 7 adult patients with scimitar syndrome, examined at the Instituto Nacional de Cardiología Ignacio Chávez from 1985 to June 2004.

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TABLE 1. Demographic Distribution of the Patients*

Case	Age at Diagnosis and Current Age, Years	Evolution Time, Years	Sex	Symptoms	Functional Class (NYHA)	Scimitar Sign on Radiography
1	6-30	24	Female	None	I	No
2	3-20	17	Female	Respiratory tract infection	II	No
3	14-28	14	Female	Respiratory tract infection	II	Yes
4	10-18	8	Female	Respiratory tract infection	II	Yes
5	17-41	24	Female	Respiratory tract infection	II	Yes
6	21-24	3	Female		II	Yes
7	27-46	19	Female	Respiratory tract infection	II	No

*NYHA indicates New York Heart Association.

PATIENTS AND METHODS

This retrospective study included 7 women with a diagnosis of scimitar syndrome examined in an adult congenital heart disease clinic. A complete clinical history, chest x-ray, transthoracic and transesophageal echocardiography, as well as left and right cardiac catheterization were performed in all patients. In 2 patients, computed tomography scanning was done to determine the site of connection of the right pulmonary veins and exclude the possibility of bronchiectasis or pulmonary hypoplasia. Three patients underwent magnetic resonance imaging, in 2 cases to define the type of connection and in 1 case to determine the site of obstruction in a tube placed during surgery.

The echocardiographic study was performed with Philips Sonos 1000 and 5500 systems, using a 2.5-mHz transthoracic probe, and biplane and multiplane transesophageal probes. Previously described specific and non-specific echocardiographic features were used for the analysis.^{4,6} The diameters of the pulmonary artery and branches were determined in the parasternal slice in both arteries. Apical and subcostal four-chamber sections obtained by two-dimensional transthoracic echocardiography were used to assess the characteristics of the atrial septal defect, pulmonary vein connections, inferior vena cava drainage, presence of obstruction in the venous collector, and dilation of the right chambers, and to detect aortopulmonary collateral vessels. Using

continuous wave color Doppler in an apical four-chamber view, the sample volume was placed at the height of the tricuspid valve and the pressure gradient between the right atrium and ventricle was determined using Bernoulli's method. The result was multiplied by a correction factor of 1.23 to obtain the pulmonary artery systolic pressure.

With the use of a four-chamber transesophageal technique, the size and location of the atrial septal defect, the manner in which the pulmonary veins were connected, and the repercussion on the right chambers were assessed. The pulmonary artery diameter was measured in the 70° longitudinal section and the pulmonary branches in the 0° section. Arterial hypoplasia was defined as a diameter of <18 mm for the pulmonary artery and <10 mm for the pulmonary artery branches.⁶

RESULTS

The patients' demographic characteristics are shown in Table 1. The most frequent symptoms were respiratory manifestations (chronic cough, decreased exercise tolerance, to exercise, and recurrent respiratory tract infection) and progressive dyspnea on heavy and moderate exertion in 90% of the cases. The scimitar sign was seen on the chest x-rays in 4 patients (57%) (Figure 1 A and B).

The transthoracic and transesophageal echocardiographic studies allowed the diagnosis to be established in 86% of the cases. The cardiac anomalies encountered are shown in Table 2. Five patients had dextrocardia, 5 dilation of the right chambers and hypoplasia of the pulmonary arteries and branches, and 5 "blunting" of the right side of the atrium (absence of right pulmonary veins draining to the left atrium).

The site of the pulmonary venous connection was determined by color Doppler: in 2 patients it was found in the lower portion of the right atrium, in 2 others in the inferior vena cava through a venous collector, and in the remaining 3 patients at the junction of the right atrium with the inferior vena cava. Three patients showed narrowing of the collector at the site where it connected with the inferior vena cava (Figure 2 A-D) (Table 3).

TABLE 2. Associated Malformations and Corrective Surgery*

Age, Years	Associated Malformation	First Surgery	Repeat Surgery
6-30	Small ASD	No	No
3-20	Large ASD	Yes	No
14-28	PDA-AoCo	Yes	No
10-18	PDA	Yes	No
17-41	ASD-PDA	Yes	Yes
21-24	Small ASD	Yes	No
27-46	Large ASD	Yes	No

*ASD indicates atrial septal defect; AoCo, aortic coarctation; PDA, patent ductus arteriosus.

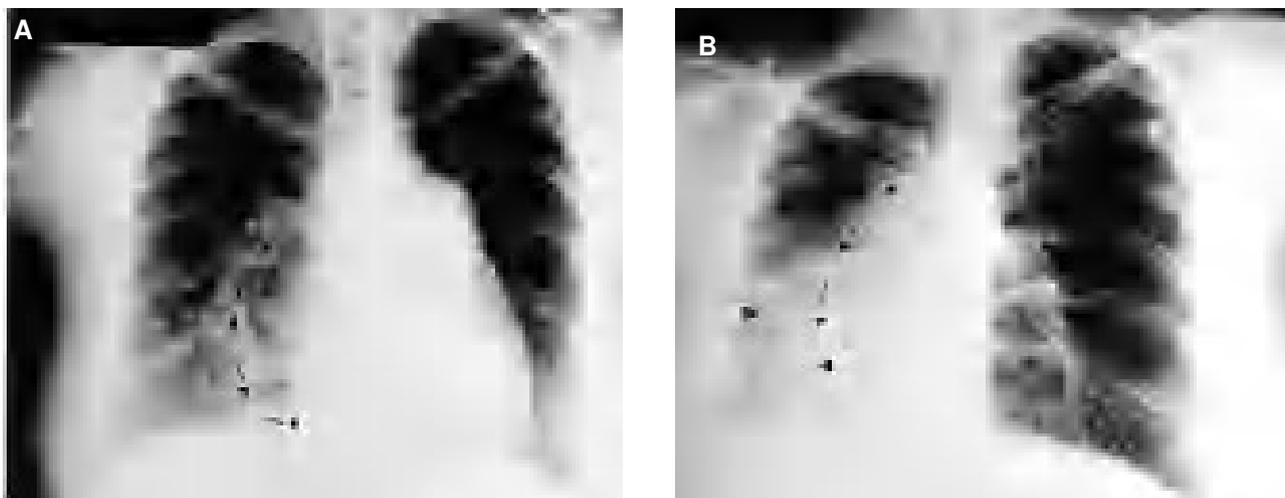


Figure 1. A: anteroposterior chest x-ray showing curved course of the collector (arrows), grade II cardiomegaly with bulging of the right pulmonary artery branch and mild hypoplasia of the lower lobe of the right lung. B: chest plain film showing dextrorotation of the heart and hypoplasia of the lower and middle lobes of the right lung. Arrows show the curved course of the collector. Arrowhead indicates an aortopulmonary collateral vessel.

The echocardiographic diagnosis showed close agreement with the findings obtained by cardiac catheterization (Figure 3), and catheterization allowed assessment of the aortopulmonary collateral circulation observed in one case.

Six patients underwent surgery to correct the associated cardiac anomaly and place a bypass from the right pulmonary veins to the left atrium. In only one of these patients, reimplantation of the right pulmonary veins to the left atrium was performed in a second

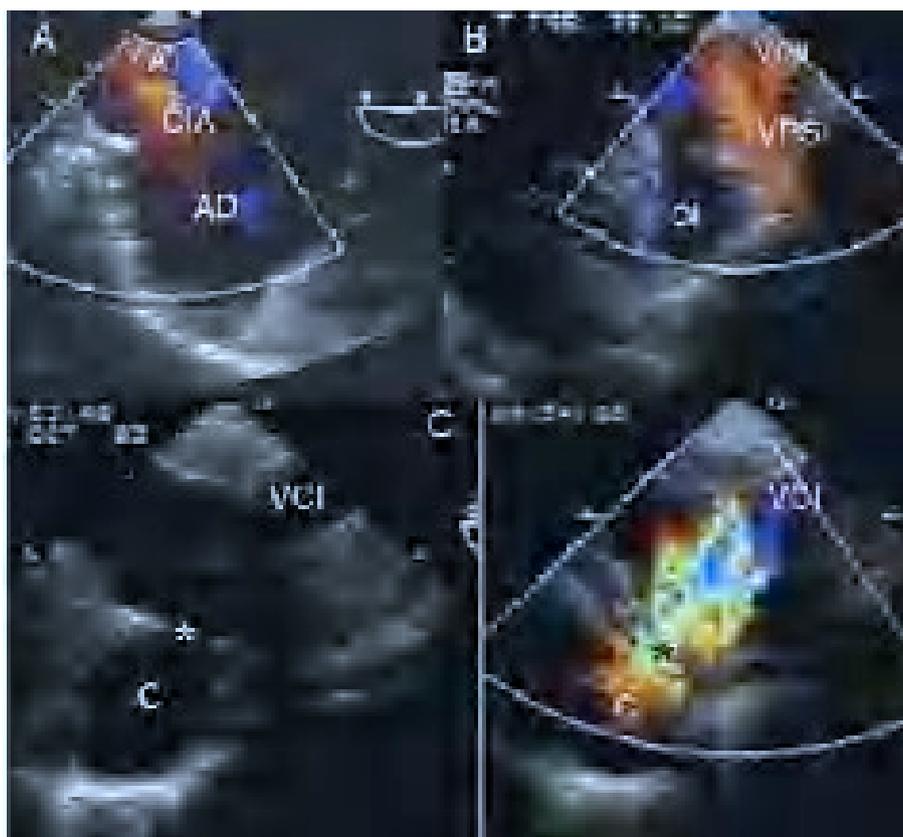


Figure 2. A: transesophageal echocardiogram with color Doppler showing the interatrial shunt. B: transesophageal color Doppler echocardiogram depicting the left pulmonary veins draining to the left atrium above the auricular appendage. C: two-dimensional and color Doppler images show turbulent flow in the collector, which drains at the site of the junction of the inferior vena cava and the right atrium.
 RA indicates right atrium; LA, left atrium; C, collector; IAC, interatrial communication; RD, right ventricle; LV, left ventricle; LAA, left auricular appendage; IVC, inferior vena cava; PV, pulmonary vein; ILPV, inferior left pulmonary vein; SLPV, superior left pulmonary vein.
 *Site of collector obstruction.

TABLE 3. Non-Specific and Specific Echocardiographic Features*

Age, Years	Dextrocardia	Dilatation of Right Chambers	Blunt Contour	Pulmonary Artery Hypoplasia	Flow to Lower Portion RA, Near IVC	Flow to RA-IVC junction	Flow to IVC
6-30	Yes	No	?	No	Yes		
3-20	Yes	No	Yes	Yes		Yes†	
14-28	No	Yes	Yes	Yes		Yes†	
10-18	Yes	Yes	Yes	?			Yes†
17-41	Yes	Yes	?	Yes			Yes
21-24	No	Yes	Yes	Yes		Yes	
27-46	Yes	Yes	Yes	Yes	Yes		

*RA indicates right atrium; IVC, inferior vena cava.

†Associated with collector obstruction.

surgical stage. One patient was reoperated for significant obstruction of the tube placed for the reimplantation.

The remaining patient had a small atrial septal defect and was in NYHA functional class I; hence, surgery was not performed (Table 2).

All the patients who underwent surgery were in functional class I at the time of writing.

DISCUSSION

Scimitar syndrome is divided into 3 groups according to the age of the patients, as defined in a multicenter study involving 122 patients aged 1 to 58 years. Group I comprises the adult form, which shows no pulmonary artery hypertension (PAH) and involves a small septal defect; it is the best tolerated, as was seen in one of our patients. Group II is characterized by an association with complex congenital abnormalities that modify the symptoms and natural history of the syndrome; the remaining patients in our series fell into this category. Group III is the infantile form and is characterized by severe PAH and a poor prognosis.^{2,4,5,7}

The most common symptoms, respiratory manifestations and progressive dyspnea,^{4,5,8} were present in all our patients. In some cases the scimitar sign is not observed because of cardiac dextrorotation or because the venous collector is not wide or curved, but instead, straight, thin or multiple. This sign presents in 70%, generally because right lung hypoplasia is absent in adults.^{1,2}

The echocardiographic diagnosis correlated well with the cardiac catheterization findings. Transthoracic echocardiography has greater value in children with scimitar syndrome, whereas in adults transesophageal echocardiography should also be performed to assess the posterior structures, such as the atrial septum to determine the size, number, morphology, and location of defects as well as the pulmonary vein connections, which cannot be well-assessed with the transthoracic technique. Transesophageal echocardiography is also useful for detecting obstruction of the tubes placed for reimplantation, as occurred in one of the cases studied, which was confirmed by magnetic resonance

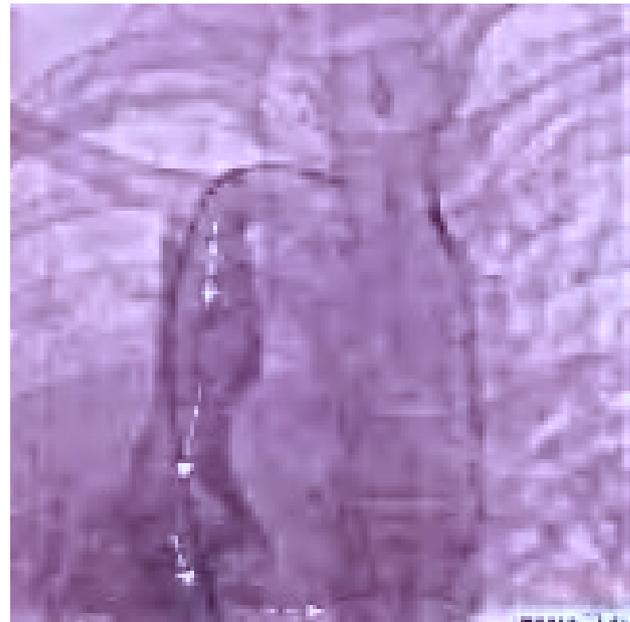


Figure 3. Angiographic study showing the curved course of the collector (arrows).

imaging.^{2,4,6,7} It is very important to determine the subdiaphragmatic areas of connection for better surgical treatment. A surgical alternative would be to reimplant the collector at the posterior wall of the left atrium with off-pump circulation.^{5,8-10}

On the basis of our study, we conclude that scimitar syndrome is rare and the diagnosis is established by echocardiography in most cases. With the development of non-invasive techniques,^{11,12} the diagnosis is complemented by cardiac catheterization only in some cases.

REFERENCES

1. Mohamed T, Idris FRCP. Diagnostic aid of transesophageal echocardiography in an adult case of scimitar syndrome: confirmation of the findings at surgery and review of the literature. *J Am Soc Echocardiogr.* 1998;11:387-92.

2. Geggel RL. Scimitar syndrome associated with partial anomalous pulmonary venous connection at the supracardiac, cardiac and infracardiac levels. *Pediatr Cardiol.* 1993;14:234-7.
3. Mardini MK, Sakati HA, Lewall DB, Cristie R, Nyhan WL. Scimitar syndrome. *Clin Pediatr.* 1982;21:350-4.
4. Dupuis C, Charaf LAC, Breviere GM, Abou P, Remy-Jardín M, Helmius G. The "adult" form of the scimitar syndrome. *Am J Cardiol.* 1992;70:502-7.
5. Salazar J. Scimitar syndrome: five cases examined with two-dimensional and Doppler echocardiography. *Pediatr Cardiol.* 1995; 16:283-6.
6. Shibuya K, Smallhorn JE, McCrindle BW. Echocardiographic clues and accuracy in the diagnosis of scimitar syndrome. *J Am Soc Echocardiogr.* 1996;9:174-81.
7. Dupuis C, Charaf L, Breviere GM, Abou P. "Infantile" form of the scimitar syndrome with pulmonary hypertension. *Am J Cardiol.* 1993;71:1326-30.
8. Cobanoglu A, Hanlon JT, Combs DT, Starr A. Combined intracardiac and extracardiac repair of scimitar syndrome with anomalous pulmonary veins to both cavae. *J Thorac Cardiovasc Surg.* 1984;88:142-9.
9. Neill CA, Ferencz C, Sabinston DC, Sheldon H. The familial occurrence of hypoplastic right lung with systemic arterial supply and venous drainage Scimitar syndrome. *Bull Johns Hopkins Hospital.* 1960;107:1-21.
10. Brown JW, Ruzmetov M, Minnich D, Vijay P, Edwards CA, Uhlig PN, et al. Surgical management of scimitar syndrome: an alternative approach. *J Thorac Cardiovasc Surg.* 2003;125:238-45.
11. Inoue T, Ichihara M, Uchida T, Sakai Y. Three-dimensional computed tomography showing partial pulmonary venous connection complicated by the scimitar syndrome. *Circulation.* 2002;105: 663.
12. Rose C, Vosschenrich R. Incomplete scimitar syndrome. *Cardiol Young.* 2002;12:389-90.