Survival and Prognostic Factors in Patients With an Absent Atrioventricular Connection

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Introduction and objectives. To identify anatomical and functional characteristics associated with survival in adult patients with an absent atrioventricular connection and to highlight the diagnostic importance of echocardiography.

Methods. The clinical histories and echocardiographic and hemodynamic test results of 24 patients were recorded.

Results. Some 87.5% of patients were in New York Heart Association (NYHA) functional class I/II. In 92%, the ECG demonstrated sinus rhythm and left ventricular dilatation. Chest x-ray showed grade-II cardiomegaly in 83%. Situs solitus and an absent right atrioventricular connection were found on echocardiography in 92%. The ventriculoarterial connection was most frequently concordant (in 71%). All patients had an atrial septal defect, 21 had a ventricular septal defect, and 21 had decreased pulmonary flow. The ejection fraction of the main ventricle in the whole patient group was 55% (10%); 52% (12%) in those who did not undergo surgery and 58% (8%) in those who did (*P*=NS). Factors associated with poor survival were an ostium secundum atrial septal defect, hemoglobin <16 g/dL, and a main ventricle ejection fraction <50%. Of the 54% of patients who underwent surgery, 85% are alive and the majority are in NYHA functional class I/II. Among those who did not, 82% are alive and 73% are in NYHA functional class I/II.

Conclusions. The presence of a wide atrial septal defect, a normal hemoglobin level, and a normal main ventricle ejection fraction were associated with the survival of these patients into adulthood. Echocardiography can provide clinicians and surgeons with information that is valuable for selecting treatment and monitoring follow-up.

Key words: *Absent atrioventricular connection. Tricuspid atresia. Adults.*

Supervivencia y factores pronósticos en la ausencia de conexión auriculoventricular

Introducción y objetivos. Analizar las características anatómicas y funcionales que favorecieron la supervivencia de pacientes adultos con ausencia de conexión auriculoventricular y resaltar la importancia del diagnóstico ecocardiográfico.

Métodos. Se estudió a 24 pacientes mediante historia clínica, ecocardiograma y estudio hemodinámico.

Resultados. La clase funcional del grupo estudiado fue I/II en el 87,5%. El electrocardiograma mostró ritmo sinusal en el 92% y crecimiento ventricular izquierdo. La radiografía de tórax demostró cardiomegalia grado II en el 83% de los casos. El ecocardiograma mostró situs solitus y ausencia de conexión auriculoventricular derecha en el 92% de los casos. La conexión ventriculoarterial concordante fue la más frecuente (71%). Todos tuvieron comunicación interauricular; 21, comunicación interventricular, y 21, alteración del flujo pulmonar. La fracción de eyección del ventrículo principal (FEVP) del grupo total fue del 55% ± 10%; en los no operados, del 52% ± 12% y en los operados, del 58% ± 8% (diferencias sin significación estadística). Los factores que disminuyen la supervivencia son: comunicación interauricular ostium secundum, cifras de hemoglobina < 16 g/dl y la FEVP < 50%. El 54% de los pacientes recibió tratamiento quirúrgico, el 85% vive y la mayoría está en clase funcional I/II. De los no operados, el 82% vive y el 73% está en clase funcional I/II.

Conclusiones. La comunicación interauricular amplia, la hemoglobina normal y la función sistólica del ventrículo principal normal fueron los factores que favorecieron la supervivencia de estos pacientes hasta la edad adulta. La ecocardiografía proporciona valiosa información al clínico y al cirujano para decidir las diferentes opciones terapéuticas y evaluar el seguimiento.

Palabras clave: *Ausencia de conexión auriculoventricular. Atresia tricuspídea. Adultos.*

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INTRODUCTION

An absence of right atrioventricular connection, a condition also known as classic tricuspid atresia, is a cyanotic congenital heart disease conferring the physiology of a univentricular heart. Associated survival is very poor and mainly depends on the balance between the systemic and pulmonary circulation. When this balance is achieved, affected patients can reach adulthood.[1] Patients with this condition can be divided into 2 main groups: those with absent right atrioventricular connection (Figure 1) and those with absent left connection (Figure 2). In hearts with absent right connection, there is no continuity between the right chambers, and the only connection is between the left atrium and left ventricle. The right atrium has a muscular floor and there is no tricuspid valve. Externally, a deep groove is seen between the right atrium and the right ventricle, which is missing the inlet and is generally underdeveloped. The left chambers are connected through the mitral valve, and the hypertrophic, dilated left ventricle acts as the main ventricle. The 2 ventricular chambers are connected by a subinfundibular ventricular septal defect. This usually presents an atrial septal defect similar to an ostium secundum, although there may be a patent foramen ovale or a common atrium. This basic morphological pattern can present several variants in individual cases, determined by the type of ventriculoarterial connection, size of the septal defects, and morphology of the cardiac valves (straddling mitral valve).

In hearts with absent left connection, the ventricles are inverted. The morphologic right ventricle positioned at the left does not maintain anatomic continuity with the left ventricle; it is the morphologic left ventricle located at the right that connects with the right atrium. The morphological variants of this condition are based on the same parameters as those mentioned above for absence of right atrioventricular connection.

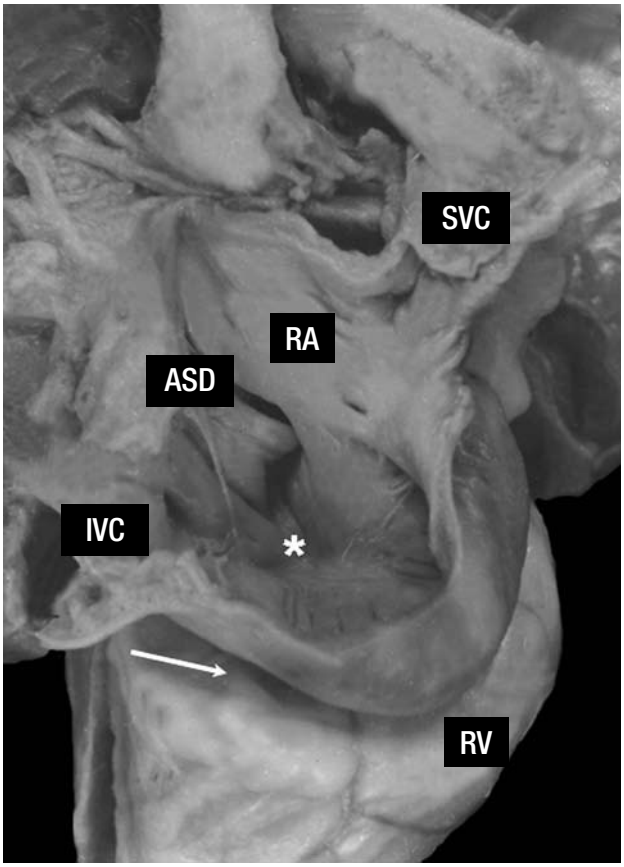

Figure 1. Interior view of the right atrium, showing absence of the atrioventricular connection. Observe the muscular floor of the atrium (asterisk) and the deep groove separating the atrium from the ventricle (arrow) (archived anatomic specimen). ASD indicates atrial septal defect; IVC, inferior vena cava; RA, right atrium; RV, right ventricle; SVC, superior vena cava.

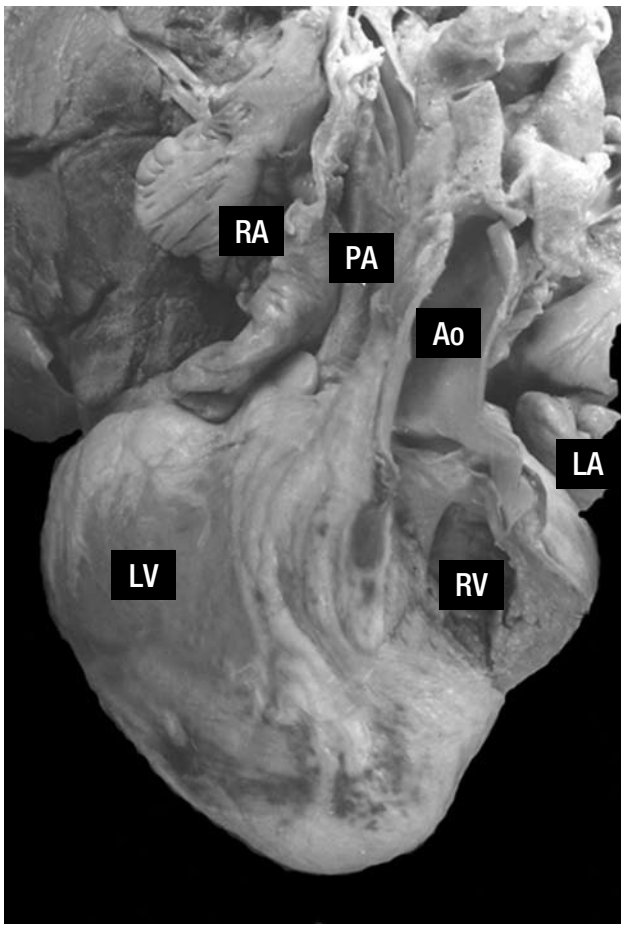

Figure 2. Heart showing absence of a left atrioventricular connection. Observe inversion of the ventricles and the discordant ventriculoarterial connection (archived anatomical specimen). Ao indicates aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle.

The ventricle that receives the atrioventricular connection manages the blood volume from both the systemic and pulmonary circulation and, therefore, it hypertrophies and dilates, whereas the ventricle without connection receives flow from a ventricular septal defect, whose size determines the degree to which this ventricle develops. There is also an obligatory atrial septal defect to derive blood from the right atrium to the left atrium, or from the left to the right in hearts with absent connection between the left atrium and the morphologic right ventricle (when the ventricles are inverted), where the systemic and pulmonary flows mix.[2,3]

In hearts with absent right atrioventricular connection, the ventriculoarterial connection is generally concordant, with the great arteries emerging from their respective ventricles. Less often, the ventriculoarterial connection is discordant, with the aorta emerging from the right ventricle and the pulmonary artery from the left ventricle. Even less commonly, the connection may be a double ventricular outlet, and in rare cases, there may be a single outlet through the common trunk or a solitary aortic trunk. In hearts with absent left atrioventricular connection, a discordant ventriculoarterial connection is most common. Some patients progress with a certain degree of pulmonary stenosis, which protects the pulmonary vascular bed from hyperflow.[2,4,5]

In unoperated patients with this heart disease, survival above age 15 is rare and complications due to dysfunction of the main ventricle are common.[6] Echocardiography is very useful to precisely determine the anatomic and functional features of this complex congenital heart disease.[7,9]

This study analyzes the clinical and morphologic characteristics that favor survival in adult patients with absence of a right or left atrioventricular connection and emphasizes the importance of echocardiographic diagnosis and follow-up, which enables the functional and anatomic aspects of this congenital heart condition to be shown in detail.

METHODS

Between January 1997 and January 2007, 24 adult patients with clinically suspected tricuspid atresia were studied, including 10 men (40%) and 14 women (60%), with a mean age of 28 (8) (range, 20-54) years. All patients underwent a clinical history, hemogram (normal hemoglobin ≤16 g/dL), arterial oxygen saturation testing (desirable, >80%), 12-lead surface electrocardiography, chest x-ray, and echocardiography; 24-hour Holter monitoring was additionally performed in all patients with a history of palpitations. In 67% of cases, cardiac catheterization was performed along with echocardiography.

Echocardiography was carried out with a Phillips Sonos 5500 unit, equipped with an S4 transducer and M-mode 2-dimensional Doppler. Sequential segmental analysis was used to characterize the heart disease.

The end-diastolic and end-systolic diameters of the main ventricle were determined by M mode in the parasternal long-axis view. The ejection fraction (EF) of the main ventricle was determined in the 4-chamber apical view, using the modified Simpson method.

Statistical Analysis

A descriptive analysis was made of the qualitative and quantitative variables with measurement of the central tendency by the mean, median, or percentage, according to the distribution of each one. Multivariate analysis with ANOVA was used for heterogeneous variances. Variables with a nonparametric distribution were analyzed with the Mann-Whitney *U* test. The survival analysis was performed with the Kaplan-Meier method and Cox regression, adjusted according to time and the following covariates: pulmonary stenosis, atrioventricular valve failure, preoperative pulmonary hypertension, hemoglobin level, atrial oxygen saturation, supraventricular arrhythmias, and ostium secundum atrial septal defect. Significance was set at a *P* value of ≤.05. SPSS version 14 was used for the analyses.

RESULTS

Clinical Manifestations

The mean time interval between birth and the diagnosis of absent atrioventricular connection was 13.8 (10) years (range, 3-23). Cyanosis was present in all patients, occurring since the first year of life in 23 (96%) patients and after the age of 6 years in 1 (4%); 21 patients (87.5%) presented acropachy.

Nearly all the patients (21/24; 87.5%) had some type of heart murmur. Among the total, 87.5% were in functional class I/II and 22.5%, in functional class III. Mean hemoglobin level was 18.2 (3.4) g/dL. Mean arterial oxygen saturation was 73% (16%).

The surface electrocardiogram showed sinus rhythm in 21 (87%) patients, atrial fibrillation in 13 (13%), significant arrhythmia in 8 (33.3%), episodes of supraventricular tachycardia in 6, and frequent atrial extrasystoles in 2. All patients had evidence of left ventricular enlargement.

On chest radiography, all patients presented some degree of cardiomegaly, including 79% (19/24) with grade II. In addition, 7 (29%) patients had evidence

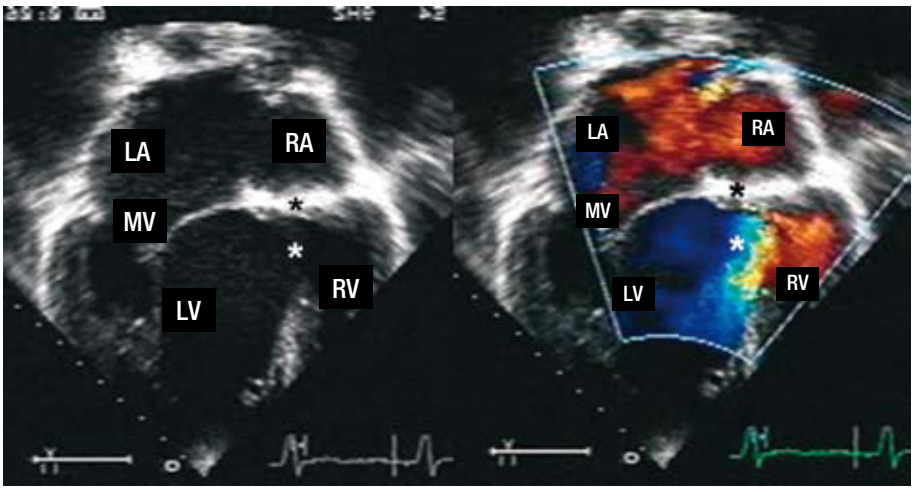

Figure 3. Echocardiographic images in a 4-chamber apical view showing absence of a right atrioventricular connection, in 2-dimensional imaging and with color Doppler. Note the echogenic area separating the right chambers (black asterisk) and the ventricular septal defect (white asterisk). Ao indicates aorta; LA, left atrium; LV, left ventricle; MV, mitral valve; RA, right atrium; RV, right ventricle.

of pulmonary obstruction, 3 normal pulmonary flow, and 14 pulmonary hypovascularization.

Surgical treatment was undertaken in 54% of patients (13/24): Blalock-Taussig shunt in 8 (61%), pulmonary artery banding in 2 (16%), and total cavopulmonary bypass in 3 (23%). Among the operated patients, 85% (11/13) are alive, 10 in functional class I/II and 1 in functional class III. Two patients in this group died (Table 1). The mean follow-up time after surgery was 13 (8) years (range, 1-28). Among the patients who did not undergo surgery, 82% (9/11) are alive, 8 in functional class I/II and 1 in functional class III. Two patients (18%) died (Table 1).

Medical treatment was given to 75% of patients (18/24); 2 of them had also undergone surgery (Table 1).

One 38-year-old woman had a spontaneous abortion, and another 25-year-old patient had a pregnancy that reached term. One patient presented systemic arterial hypertension, and the pertinent tests demonstrated the additional presence of Takayasu arteritis. Three patients had a cerebral infarction, 2 due to infectious endocarditis and cerebral embolism, and 1 due to neurocysticercosis.

Echocardiography Characteristics

Echocardiography showed the following features: 92% of cases presented situs solitus and 8%, right atrial isomerism; 88% of hearts were in levocardia and 12%, in dextrocardia; 92% presented absence of right atrioventricular connection (Figure 3) and 8% absence of left atrioventricular connection and ventricular inversion (Figure 4).

The ventriculoarterial connection was concordant in 71% of patients (Figure 5) and discordant in 17%; 12% had a double outlet.

All patients had an atrial septal defect, with a mean diameter of 27.6 (12) mm; 20 (83%) had an ostium secundum defect, with a mean diameter of 23.3 (7.4) mm, and 4 (17%) had a common atrium type, with a mean diameter of 48.7 (5.6) mm. In 19 (79%) patients, a subinfundibular (muscular) ventricular

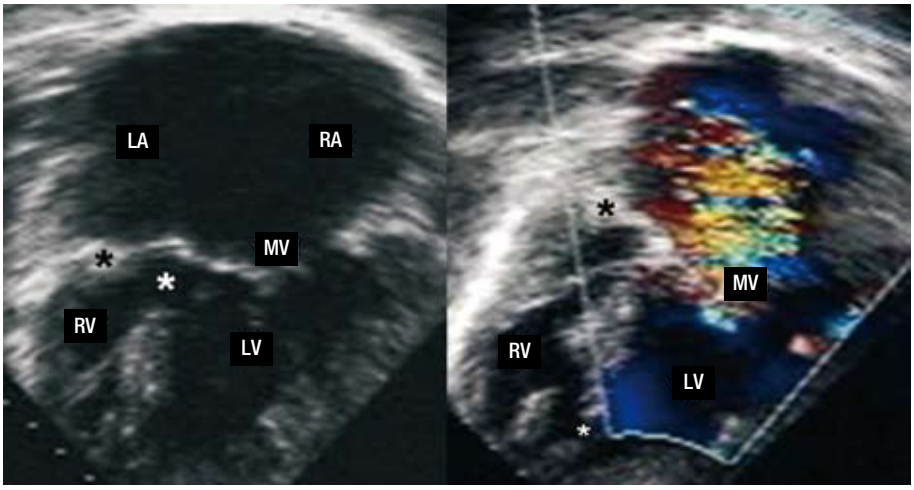

Figure 4. Two-dimensional echocardiographic image in a 4-chamber apical view showing absence of the left atrioventricular connection (black asterisk) with ventricular inversion. Two ventricular septal defects are observed (white asterisk). The color Doppler image depicts severe mitral regurgitation. LA indicates left atrium; LV, left ventricle; MV, mitral valve; RA, right atrium; RV, right ventricle.

TABLE 1. **Clinical Data of the Patients (n=24)**

Patient	Age, y	Sex	Hemoglobin, g/dL	PO_2,%	Arrhythmia	Surgery	Medical Treatment	NYHA Functional Class	Alive
1	21	Man	16.6	54.2	No	No	No	I	Yes
2	21	Man	21.1	38	SVT, WPW	No	Digoxin, diuretic, O_2, NTG	III	No
3	22	Man	23	68.1	No	BT shunt	Digoxin, enalapril, Sintrom	I	Yes
4	30	Man	18.4	83	No	BT shunt	Aspirin	I	Yes
5	33	Man	17.1	75	SVT, VT, WPW, AF	No	Propafenone, Sintrom	II	No
6	54	Woman	18.4	70	No	No	Diuretic, Sintrom, O_2, nifedipine	II	Yes
7	30	Woman	21	76	No	No	Diuretic, aspirin, pentoxifylline	III	Yes
8	38	Woman	20.5	80.2	No	No	Diltiazem, aspirin	I	Yes
9	23	Woman	21	73	No	No	No	II	Yes
10	21	Woman	19.2	63	No	Fontan	No	I	Yes
11	36	Woman	14.9	90	No	BT shunt	Digoxin, diuretic, Sintrom, captopril	III	Yes
12	22	Man	14.2	88	No	BT shunt	Digoxin, diuretic, aspirin	I	Yes
13	20	Man	15.9	86	SVT	No	No	II	Yes
14	25	Woman	11	48	Atrial extrasystoles	Pulmonary banding	No	II	Yes
15	27	Man	17.1	76	No	No	Sintrom	II	Yes
16	23	Woman	15.2	88	No	Fontan	Diuretic, amlodipine	II	Yes
17	37	Woman	16.9	40	SVT, AF, BAV-II	BT shunt	Digoxin, diuretic, atenolol, pacemaker	I	Yes
18	24	Woman	15.4	78	No	No	Aspirin, propranolol	II	Yes
19	30	Woman	14.7	78	SVT, AF	BT shunt	Digoxin, diuretic, Sintrom, enalapril, carvedilol	II	Yes
20	20	Man	24.2	69	Atrial extrasystoles	Pulmonary banding	Digoxin, diuretic, aspirin	II	No
21	30	Woman	15.6	88	No	BT shunt	Digoxin, diuretic	I	Yes
22	37	Woman	21.8	70.7	No	Fontan	Aspirin	II	No
23	28	Man	20.9	89.3	No	No	No	II	Yes
24	22	Woman	23.6	90	SVT	BT shunt	Digoxin, diuretic, O_2	II	Yes
Mean (SD)	28 (8)	Men,10; women, 14	18.2 (3.4)	73 (16)	Arrhythmia, 8	Operated, 13	Treatment, 18		

AF indicates atrial fibrillation; BT, Blalock-Taussig; NTG, nitroglycerin; SVT, supraventricular tachycardia; VT, ventricular tachycardia; WPW, Wolff-Parkinson-White syndrome.

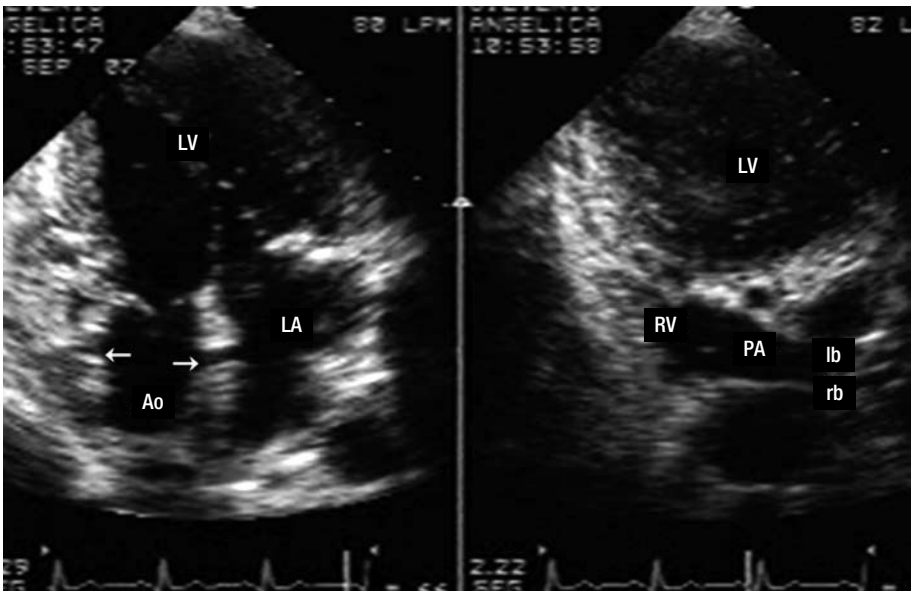

Figure 5. Two-dimensional echocardiographic image in a 4-chamber apical view showing a concordant ventriculoarterial connection (the arrows indicate emergence of the coronary arteries). Ao indicates aorta; ASD, atrial septal defect; IVC, inferior vena cava; LA, left atrium; lb, left branch; LV, left ventricle; PA, pulmonary artery; RA, right atrium; rb, right branch; RV, right ventricle; SVC, superior vena cava.

defect was found, and 3 of these patients also had a trabecular (muscular) ventricular septal defect. In another 3 (12%) patients, the ventricular septal defect was obliterated. In the 2 patients with absence of left atrioventricular connection, the ventricular septal defect was an infundibular subarterial variety (musculomembranous).

The mean diameter of the ventricular septal defect was 13.9 (7.4) mm. In the above-mentioned 2 patients, in particular, who are interesting and uncommon, the atrial septal defect was restrictive in 1 and wide in the other, and the ventriculoarterial connection was discordant in 1 and a double outlet of the main ventricle in the other; both patients had pulmonary stenosis and 1 of them presented right-sided isomerism.

Pulmonary flow was abnormal in 21 (87.5%) patients: 6 had pulmonary valve stenosis, 4 mixed (valvular and subvalvular) pulmonary stenosis, and 2 pulmonary subvalvular stenosis. Five had pulmonary hypoplasia, 4 pulmonary atresia, and 3 (12.5%) patients showed no pulmonary stenosis (Table 2).

The concomitant anomalies included Chiari network (2 patients), persistent ductus arteriosus (2 patients), and persistent left superior vena cava to the coronary sinus (1 patient).

In the overall group, mean left ventricular diastolic diameter was 61 (11) mm, mean right ventricular diameter was 16 (7) mm; and mean EF of the main ventricle was 55% (10%). When patients were divided into nonoperated and operated groups, EF of the main ventricle was 52% (12%) and 58% (8%), respectively (statistically nonsignificant differences).

Mitral failure, documented in 17 patients, was mild in 7 (41%) and moderate or severe in 10 (59%) (Table 2).

In the Kaplan-Meier survival analysis and Cox regression with relative risk function, the variables associated with decreased survival in these patients were presence of an ostium secundum atrial septal defect with respect to a common atrium, which was statistically significant by log rank test ($P<.01$) (Figure 6), and hemoglobin values >16 g/dL, which showed a trend to significance ($P=.08$).

The mean survival of patients since a decrease in the EF of the main ventricle was detected (<50%) was 12 years after adjusting for time and the covariates (Figure 7).

Sixteen patients (67%) underwent cardiac catheterization. Pulmonary hypertension was documented in 6 cases before surgery and in 3 cases following surgery (Table 2).

DISCUSSION

An absence of right atrioventricular connection (classic tricuspid atresia) has a broad spectrum of anatomic presentations. Survival to adulthood depends on the balance achieved between the systemic and pulmonary circulation.[10] The condition is characterized by the presence of a main ventricle that must sustain the systemic, pulmonary, and coronary circulation. When an absence of right atrioventricular connection is detected in a timely manner and other factors are favorable, patients should undergo surgery for physiologic correction of the problem, ideally using a Fontan technique.[11,12] In patients

TABLE 2. **Echocardiographic and Hemodynamic Data (n=24)**

Patient	Type of ASD	ASD Diameter, mm	MV Diastolic Diameter	EF of Main Ventricle, %	Pulmonary Flow Obstruction		AV Valve Regurgitation	Preoperative Catheterization (MPAP/LVEDP)		Postoperative Catheterization (MPAP/LVEDP)	
					Type	Maximum Gradient, mm Hg					
1	OS	16	50	60	Mixed PS	65		29	13		
2	OS	19	51	50	Valvular PS	96	Mild MR				
3	OS	13	70	55	Pulmonary atresia		Moderate MR				
4	AC	47	60	60	Mixed PS	81	Severe MR				
5	OS	23	64	25	Valvular PS	89	Mild MR				
6	AC	42	72	35			Severe MR	65	16		
7	OS	30	50	60	Pulmonary hypoplasia		Mild MR				
8	OS	36	54	60				36	12		
9	OS	20	52	60	Pulmonary hypoplasia		Mild MR				
10	OS	15	62	66	Pulmonary hypoplasia		Mild MR			9	2
11	OS	21	45	47	Mixed PS	91	Moderate MR			20	
12	OS	29	45	55	Pulmonary atresia			42	12		
13	OS	33	76	57	Mild MR			60	7		
14	OS	35	73	52	Subvalvular PS	82		30	14		
15	OS	20	51	60	Pulmonary hypoplasia		Moderate MR				
16	OS	12	51	70	Pulmonary atresia						
17	OS	16	84	56	Pulmonary atresia		Moderate MR			46	18
18	OS	22	72	55	Mixed PS	32	Moderate MR	18	7		
19	AC	55	65	50	Valvular PS	54	Moderate MR	16	13	30	21
20	AC	51	81	60	Subvalvular PS	81		12		LB	42
21	OS	20	55	68	Pulmonary hypoplasia			12	6		
22	OS	27	69	64	Valvular PS	88		18	15		
23	OS	31	53	48	Valvular PS	43	Mild MR	18	8		
24	OS	29	59	45	Valvular PS	85	Severe MR	9			
Mean (SD)		27 (12)	61(11)	55 (10)							

AV indicates atrioventricular; ASD, atrial septal defect; CA, common atrium; LB, left branch of pulmonary artery; LVEDP, left ventricular end-diastolic pressure; MPAP, mean pulmonary artery pressure; MR, mitral regurgitation; MV, main ventricle; MVEF, main ventricle ejection fraction; OS, ostium secundum; PS, pulmonary stenosis; SD, standard deviation.

with a restrictive atrial septal defect, the systemic venous pressure is increased.

Patients with an absence of left atrioventricular connection with ventricular inversion have different hemodynamics, because the pulmonary venous return is affected, with a consequent elevation of venous and capillary pressure. Despite the problems associated with this condition, our 2 patients survived to adulthood; 1 was treated with a systemic-pulmonary shunt, and the other did not undergo surgery. The abovementioned factors indicate that the 2 types of absent connections present different hemodynamic patterns.

In our setting, some patients receive a late diagnosis and others do not meet the requisites for Fontan surgery, whereas other patients are opportunely diagnosed and undergo surgery.[13-16] This study includes 24 patients who survived to more than 20 years of age, are independent, and carry out simple tasks, in general with a good functional class. Thirteen patients underwent some type of surgical intervention that allowed survival to this

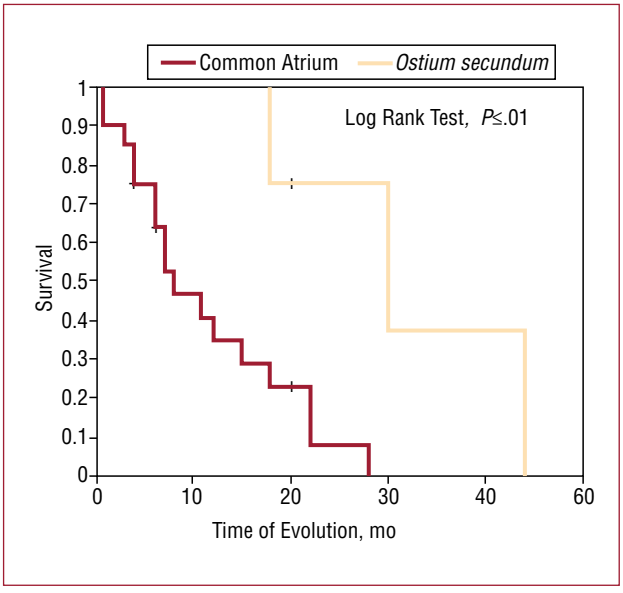

Fig. 6. Tricuspid atresia. Patient survival as related to the size of the atrial septal defect. Statistically significant difference with the log rank test ($P \leq .01$).

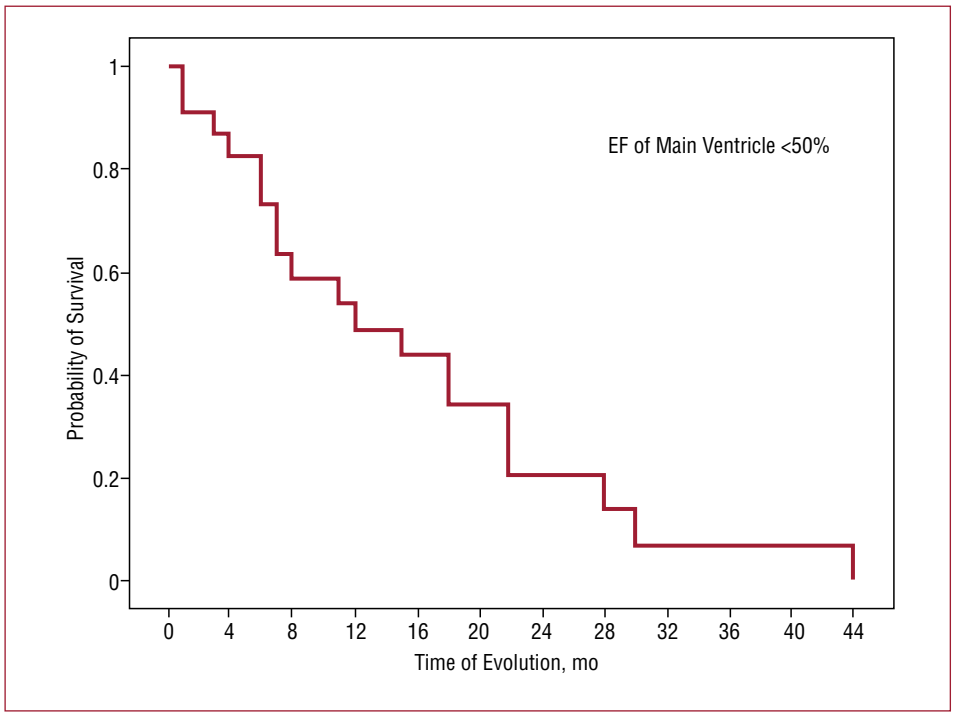

Figure 7. Mean survival of patients from the moment at which a decrease in the ejection fraction of the main ventricle is detected (<50%), adjusted by time and covariates.

age. However, as would be expected considering the natural evolution of this condition, there were 4 deaths: 1 patient who had undergone pulmonary banding, 1 treated with Fontan surgery, and 2 who were not surgically treated.

Pregnancy is not recommended in patients with a univentricular heart physiology. In addition to the circulation that must be managed, the fetal/placental circulation implies an even greater load on the main ventricle. Nonetheless, 1 patient in our series was able to bring her pregnancy to term despite the risks involved.[17]

Three patients presented cerebrovascular events during follow-up, which were associated with septic embolisms and neurocysticercosis. These cyanotic patients have a higher risk of cerebrovascular events than the general population, and the risk increases when there is hypertension, atrial fibrillation, or a history of phlebotomy or microcytosis ($P<.05$). In contrast, there was no relationship between cerebrovascular events and the patient's age, smoking habit, erythrocytosis, ejection fraction, or treatment with aspirin or coumarin derivatives.[18] In our study the statistical analysis showed that patients with polyglobulia have a tendency to lower survival, although the difference was not statistically significant.

The echocardiographic study enabled determination of the patients' morphologic and functional characteristics. We found that although the main ventricle was dilated in all cases, systolic function was preserved both in patients who had undergone surgery and those who had not. This fact may have favored an adequate quality of life, as was indicated by their functional class. An ostium secundum atrial septal defect and decreased systolic function of the main ventricle were factors with a negative influence on survival. Most of the studies focusing on main ventricle function in this pathology have been performed in children[15,19] the present study is among the first undertaken in a series of adult patients.

Fontan surgery has been of great help to these patients; nonetheless, 46% in our series did not undergo any type of surgery and survived to adulthood.[13,16] The presence of pulmonary stenosis was essential in these patients because it protected the vascular bed from pulmonary hyperflow and consequent injury to the pulmonary vasculature, although, on the other hand, it also favored polyglobulia.

The patients who were treated with some type of surgery survived longer and in a better functional class than those who experienced the natural evolution of the condition, although these differences were only trends and were not statistically significant. The decision to administer medical treatment was based on the patient's functional class and systolic function of the main ventricle calculated by echocardiography.

The limitations of this study include the small number of patients with dysfunction of the main ventricle and/or a common atrium, and the possible modifications of the prognosis owing to medical treatment.

CONCLUSIONS

In this study, a large atrial septal defect, normal hemoglobin values, and systolic function of the main ventricle >50% were factors favoring survival to adulthood in patients with absent atrioventricular connection. Echocardiography is a highly useful noninvasive technique for the anatomic and functional diagnosis of this condition, and provides the clinician and surgeon with valuable information for establishing treatment decisions and assessing the progress of these patients during follow-up.

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