Cor triatriatum sinistrum in childhood. A single institution's experience

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INTRODUCTION: Cor triatriatum sinistrum (CTS) is a rare congenital cardiac defect that can present with a wide range of symptoms and may be associated with other structural cardiac defects. Very limited data are available for pediatric patients.

OBJECTIVE: To analyze the experience with CTS at a single pediatric centre, highlighting symptoms on presentation, mode of diagnosis and outcome.

METHODS: Hospital databases were searched to identify patients with CTS who were diagnosed between 1954 and 2005. Medical records with demographic data, clinical evaluation, diagnostic approach, interventions and autopsy results were reviewed.

RESULTS: Between 1954 and 2005, 82 patients (43 female children, 52%) with CTS were diagnosed at the institution. Patients were born between 1951 and 2004, and the median age at presentation was eight months (range one day to 16.1 years). The majority of patients (77%) presented with associated cardiac lesions. Of 82 patients, 57 (70%) underwent resection of the fibromuscular diaphragm, 14 (17%) did not require surgery and 11 (13%) did not survive the time to intervention. Nineteen patients (23%) died a median of two months (range one day to 5.5 years) after presentation. Nine of these patients (11%) died before surgery was attempted. Most patients were completely asymptomatic at the time of the last follow-up at a median of three years (range one day to 18 years). Echocardiography significantly facilitated the diagnosis and subsequent management of patients with CTS.

CONCLUSIONS: Despite the diversity in presentation and complexity of associated lesions, the outcome for patients with CTS is favourable.

Key Words: Congenital heart disease; Echocardiography; Pediatrics

Cor triatriatum sinistrum (CTS) is a rare congenital cardiac defect in which the left atrium is subdivided by a fibromuscular membrane into two distinct chambers. The defect was first described in 1868 in a 38-year-old woman (1). The pulmonary veins drain into a posteriosuperior proximal chamber, whereas the distal chamber communicates with the left atrial appendage and the mitral valve (2). Examples of embryological explanations for the development of the subdividing membrane are the malseptation, malincorporation and entrapment hypotheses (3). Based on studies on human embryos, Van Praagh and Corsini (2) support the entrapment hypothesis, suggesting that CTS results from entrapment of the left atrial ostium of the common pulmonary vein by tissue of the right horn of the sinus venosus, leading to failure of incorporation of the common pulmonary vein into the left atrium during the fifth embryonic week. However, attempts to clarify the embryogenesis are limited (2,3).

Cœur triatrial gauche chez l'enfant. Expérience d'un établissement

INTRODUCTION : Le cœur triatrial (CTA) gauche est une anomalie cardiaque congénitale rare, parfois accompagnée de toute une gamme de symptômes et associée à d'autres anomalies cardiaques structurales. On dispose de très peu de données à son sujet chez les patients pédiatriques.

OBJECTIFS : Analyser l'expérience acquise sur le CTA dans un centre pédiatrique en mettant en lumière les symptômes au moment de la consultation, le diagnostic et l'issue.

MÉTHODES : Les auteurs ont interrogé les bases de données de l'hôpital afin de recenser les patients atteints de CTA ayant reçu leur diagnostic entre 1954 et 2005. Ils ont passé en revue les dossiers médicaux comprenant les données démographiques, l'évaluation clinique, l'approche diagnostique, les interventions et les résultats de l'autopsie.

RÉSULTATS : Entre 1954 et 2005, 82 patients (43 fillettes, 52 %) ont reçu le diagnostic de CTA dans cet établissement. Les patients étaient nés entre 1951 et 2004 et l'âge médian au moment de la consultation initiale était de huit mois (entre 1 jour et 16,1 ans). La majorité des patients (77 %) présentaient des lésions cardiaques associées. Sur les 82 patients, 57 (70 %) ont subi une résection de la cloison fibromusculaire, 14 (17 %) n'ont pas eu besoin de chirurgie et 11 (13 %) n'ont pas survécu jusqu'à l'intervention. Dix-neuf patients (23 %) sont décédés après une période médiane de deux mois (entre 1 jour et 5,5 ans) suivant la consultation initiale. Neuf de ces patients (11 %) sont décédés avant que la chirurgie n'ait pu être tentée. La plupart des patients étaient entièrement asymptomatiques au moment du dernier suivi après une période médiane de trois ans (entre 1 jour et 18 ans). L'échocardiographie a grandement facilité le diagnostic et la prise en charge subséquente des patients atteints de CTA.

CONCLUSIONS : Malgré la diversité et la complexité des lésions associées, l'issue est restée favorable chez les patients atteints de CTA.

Patients often present with symptoms of obstruction to pulmonary venous return, resulting in pulmonary venous and, subsequently, pulmonary arterial hypertension. Age and symptoms at the time of diagnosis are highly variable, and are dependent on the degree of obstruction to pulmonary venous return, and the presence of associated cardiac lesions. Mild forms of CTS with a large opening can remain unrecognized; there are reports of incidental diagnosis in late adulthood (4-6).

Due to the rarity of CTS, the number of cases reported in the literature is relatively sparse and other than several case reports (7-11), there are very few case series with small participant numbers that include both pediatric and adult patients. Very limited data are available regarding residual morbidity following surgical correction and long-term follow-up in children. The present report represents a single-institution analysis of pediatric patients with CTS, with

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TABLE 1
Demographics and mode of diagnosis of patients with cor triatriatum sinistrum

	All patients (n=82)	1954–1981 (n=20)	1982–2005 (n=62)	P
Sex, male:female	39:43	9:11	30:32	0.81
Age at diagnosis, median (range)	8 months (1 day – 16.1 years)	9 months (1 day – 16.1 years)	8 months (2 days – 16 years)	0.92
Mode of diagnosis, n (%)				
Echocardiography	60 (73)	6 (30)	54 (87)	
Cardiac catheterization	11 (14)	6 (30)	5 (8)	
Surgery	2 (2)	0	2 (3)	
Autopsy	9 (11)	8 (40)	1 (2)	

TABLE 2
Symptoms and clinical findings of patients with cor triatriatum sinistrum

Symptoms/clinical findings	All patients (n=82)	1954–1981 (n=20)	1982–2005 (n=62)	P
Congestive heart failure	52 (63)	18 (90)	34 (55)	<0.01
Low cardiac output syndrome	11 (13)	6 (30)	5 (8)	0.02
Respiratory complaints	53 (65)	17 (85)	36 (58)	0.04
Failure to thrive	37 (45)	9 (45)	28 (45)	1.00
Feeding intolerance	26 (32)	4 (20)	22 (35)	0.27
Cyanosis	27 (33)	11 (55)	16 (26)	0.03
Decreased exercise tolerance	20 (24)	9 (45)	11 (18)	0.02
Palpitations, arrhythmia	3 (4)	0 (0)	3 (5)	1.00
Hepatomegaly	24 (29)	11 (55)	13 (21)	0.01
Cardiomegaly on chest x-ray	40 (49)	16 (80)	24 (39)	<0.01
Increased pulmonary vascularity on chest x-ray	21 (26)	4 (20)	17 (27)	0.57
Pulmonary edema on chest x-ray	22 (27)	10 (50)	12 (19)	0.02
Pulmonary hypertension	55 (67)	15 (75)	40 (65)	0.43
Abnormal right ventricle function	25 (30)	5 (25)	20 (32)	0.59

Data presented as n (%)

special emphasis on symptoms at presentation, mode of diagnosis and outcome.

METHODS

The databases of the Divisions of Cardiology and Cardiovascular Surgery at The Hospital for Sick Children (Toronto, Ontario) were searched to identify patients with cor triatriatum sinistrum who were diagnosed between 1954 and 2005. Medical records with demographic data and clinical evaluation, diagnostic approach (cardiac catheterization reports and echocardiograms) and interventions (surgical procedures), as well as autopsy results were reviewed.

Patients were divided into two groups based on the time of diagnosis (1954 to 1981 and 1982 to 2005) because two-dimensional echocardiography became available in 1982 as the standard of care at The Hospital for Sick Children.

Data are presented as frequencies, means $\pm$ SDs, and medians with ranges as appropriate. Differences between pre- and postechocardiography eras were assessed using Student's *t* test and Fisher's exact test, while differences between patient groups (died before surgery, discharged without surgery or underwent surgical intervention) were evaluated through ANOVA analysis and general linear regression models. Survival analysis for mortality used nonparametric Kaplan-Meier estimates. Analysis of serial echocardiography measurement z-scores over time was performed through mixed linear-regression models adjusted for repeated measures with a compound symmetry covariance structure. All statistical analyses were performed using SAS 9.1 (SAS Institute Inc, USA).

RESULTS

Patients

Between January 1954 and December 2005, CTS was diagnosed in 82 patients at The Hospital for Sick Children (Table 1). Forty-three patients (52%) were female and 39 patients (48%) were male. Patients were born between 1951 and 2004, with a

median age at presentation of eight months (range one day to 16.1 years). The median weight at the time of diagnosis was 7.0 kg (range 1.4 kg to 63.5 kg) and the median height was 70 cm (range 41 cm to 180 cm).

Originally, 63 patients (77%) were referred because of symptoms associated with a possible cardiac anomaly, while 19 (23%) were asymptomatic and referred because of incidental findings or after the detection of a heart murmur.

Eight patients (10%) were referred to the institution with the correct diagnosis of CTS. During the initial clinical assessment, an additional 54 patients (66%) were diagnosed, while the remaining 20 patients had a median delay in diagnosis of two months (range one day to 16 years). When comparing the two time periods, of the 20 patients who were referred between 1954 and 1981, 60% had a delayed diagnosis; this proportion was reduced to 13% in the 62 patients who were referred since 1982 ($P<0.01$).

From 1954 to 1981, the diagnosis was established by cardiac catheterization or autopsy in the majority of patients (70%) (Table 1). The remaining patients (30%) were diagnosed by echocardiography (M-mode), which was introduced as a novel diagnostic tool in 1978. In 1982, two-dimensional echocardiography became available, which greatly facilitated the diagnosis of structural congenital heart disease. Since then, two-dimensional echocardiography has been used for the diagnosis of 54 patients (87%). Cardiac catheterization, exploratory surgery and autopsy played a minor role for diagnosis.

Symptoms

A total of 63 patients (77%) presented with cardiac symptoms; 41 of these (65%) had symptoms most likely caused directly by the diaphragm in the left atrium, and the remaining patients had symptoms related to other cardiac anomalies. Patients presented with a very heterogeneous spectrum of symptoms that are detailed in Table 2. Patients seen before 1982 had a significantly higher occurrence of cardiac symptoms ($P=0.03$) and a significantly higher age

TABLE 3
Hemodynamic and anatomical information related to cor triatriatum sinistrum

	All patients	1954–1981	1982–2005	P
Restriction on membrane level, n/n (%)				0.01
Nonrestrictive or mildly restrictive	40/80 (50)	4/19 (21)	36/61 (59)	
Moderately to severely restrictive, imperforate	40/80 (50)	15/19 (79)	25/61 (41)	
Perforation size, mm, mean $\pm$ SD	6.44 $\pm$ 4.85*	5.50 $\pm$ 4.33 [†]	6.61 $\pm$ 4.95 [‡]	0.51
Right ventricular systolic pressure, mmHg, mean $\pm$ SD	61.37 $\pm$ 30.60 [§]	81.00 $\pm$ 25.63	54.17 $\pm$ 29.41**	0.01

*n=64; [†]n=10; [‡]n=54; [§]n=42; ^{||}n=12; **n=30

TABLE 4
Associated cardiac diagnosis

Associated cardiac diagnosis	All patients	1954–1981	1982–2005	P
Atrial septal defect	39/73 (53)	7/16 (44)	32/57 (56)	0.41
Ventricular septal defect	7/73 (10)	1/16 (6)	6/57 (11)	1.00
Anomalous pulmonary venous drainage (total or partial)	22/82 (27)	6/20 (30)	16/62 (26)	0.78
Patent ductus arteriosus	15/82 (18)	2/20 (10)	13/62 (21)	0.34
Pulmonary vein stenosis	5/82 (6)	0/20 (0)	5/62 (8)	0.33
Single ventricle	9/82 (11)	4/20 (20)	5/62 (8)	0.21
All	63/82 (77)	17/20 (85)	46/62 (74)	0.38

Data presented as n/n (%)

TABLE 5
Comparison of patients with cor triatriatum sinistrum related to intervention

	Death before intervention	Surgical resection of membrane	No intervention
Number of patients	11/82 (13)	57/82 (70)	14/82 (17)
Age at presentation, median (range)	1 week (1 day – 5 years)	8 months (1 day – 16 years)	2.5 years (12 days – 14 years)
Symptomatic at presentation	11/11 (100)	48/57 (84)	3/14 (21)
Low cardiac output syndrome	6/11 (55)	5/57 (9)	0/14 (0)
Cardiomegaly	7/11 (64)	30/57 (53)	2/14 (14)
Hepatomegaly	6/11 (55)	18/57 (32)	0/14 (0)
Pulmonary edema	6/11 (55)	16/57 (28)	0/14 (0)
Size of membrane perforation, mm, mean $\pm$ SD	2.6 $\pm$ 2.5	5.8 $\pm$ 4.0	11.4 $\pm$ 5.6
Associated cardiac diagnoses	10/11 (91)	46/57 (81)	6/14 (43)
Atrial septal defect	3/6 (50)	31/53 (58)	5/14 (36)
Ventricular septal defect	0/6 (0)	5/53 (9)	2/14 (14)
Single ventricle	5/11 (45)	4/53 (7)	0/14 (0)
PAPVD	1/11 (9)	8/57 (14)	0/14 (0)
TAPVD	4/6 (67)	9/57 (16)	0/14 (0)
Pulmonary vein stenosis	0/11 (0)	5/57 (9)	0/14 (0)
LVOTO	1/6 (17)	3/53 (6)	0/14 (0)

Data presented as n/n (%) unless otherwise indicated. LVOTO Left ventricular outflow tract obstruction; PAPVD Partial anomalous pulmonary venous drainage; TAPVD Total anomalous pulmonary venous drainage

at presentation (P=0.03). On cardiac evaluation, these patients were also in worse clinical condition. The former group presented with a higher degree of restriction through the opening of the diaphragm (P=0.01) and significantly higher right ventricular systolic pressures (P=0.01) (Table 3).

Patients who did not require corrective intervention were significantly less likely to present with symptoms (P<0.01) and presented at a significantly older age (P=0.04) than patients who underwent surgical intervention. Minor restriction (P<0.01), larger perforation size (P<0.01), low right ventricular systolic pressure (P<0.01) and the absence of cardiac symptoms at diagnosis (P<0.01) were all found to be strong predictors of being discharged without requiring surgical resection of the diaphragm.

Associated cardiac defects

The majority of patients (77%) presented with associated cardiac lesions (Table 4). Most often, CTS was found along an atrial septal defect (53%), a patent ductus arteriosus (18%), or partial or total anomalous pulmonary venous drainage (27%). No differences were

found between the two periods of diagnosis (P=0.38). Patients who did not require surgical resection of the membrane were significantly less likely to have an associated cardiac diagnosis (P=0.02) than patients who either died before resection or who required surgical intervention. Single ventricles were observed in nine patients (11%), all of whom either died before intervention or required complex surgical care (Table 5).

Associated noncardiac lesions

More than one-third of patients (35%) also presented with associated noncardiac lesions, mainly chromosomal abnormalities (12%) and respiratory issues (11%). There were no differences between periods of diagnosis (P=0.61) and between primary outcomes (P=0.70) in the distribution of associated noncardiac diagnoses.

Surgery

Of 82 patients, 57 (70%) underwent resection of the fibromuscular diaphragm, 14 patients (17%) did not require surgery and 11 (13%) did not survive the time to intervention. During the first time period,

Figure 1) Survival of patients with *cor triatriatum sinistrum* after diagnosis. Era 1 includes all patients diagnosed between 1956 and 1982, and era 2 includes all patients diagnosed between 1983 and 2005

Figure 2) Survival of patients with *cor triatriatum sinistrum* since surgery. Era 1 includes all patients diagnosed between 1956 and 1982, and era 2 includes all patients diagnosed between 1983 and 2005

11 patients underwent surgery compared with 46 patients during the second time period. Patients diagnosed during the first time period underwent significantly more cardiac catheterizations ($P=0.03$), mostly due to the fact that this was the main tool of diagnosis. A total of 44 patients (77%) had other cardiac procedures, most often atrial septal defect repair (46%) and ligation of a patent ductus arteriosus (16%). Immediate postoperative complications were observed in more than one-half (58%) of the patients who underwent membrane resection. Prolonged ventilation (21%), pulmonary hypertension (18%), acute arrhythmia (18%) and decreased cardiac function (16%) were the most commonly reported complications. No differences were found in the frequency of complications by period of diagnosis ($P=0.75$) and no reoperations were necessary.

Mortality and prognosis

A total of 19 patients (23%) died a median of two months (range one day to 5.5 years) after presentation. Early death after surgery (within 30 days) occurred in five of the patients (9%). Four deaths occurred in the first group (36% early mortality), whereas only one early death (2%) was noted in the second observation period. This was related to a complicated course with renal failure in a patient with right atrial isomerism after Fontan takedown.

Postdischarge cardiac symptoms (11%) and mild residual pulmonary hypertension (3%) were both seen infrequently on echocardiography. The majority of patients were completely asymptomatic at the time of last follow-up at a median of three years (one day to 18 years) after initial presentation at The Hospital for Sick Children.

Figure 3) Changes over time of right ventricular systolic pressure (RVSP) after surgery in patients with *cor triatriatum sinistrum*. The solid line represents the estimate of change in RVSP over time and the dashed lines represent the 95% CIs. P values are the estimate of the statistical significance of the observed time trend

Survival from the initial visit at The Hospital for Sick Children was estimated to be 84% at three months, 78% at one year and 74% at five years (Figure 1). Survival after resection or discharge (for nonsurgical patients) was found to be 94% at three months, 88% at one year and 86% at five years (Figure 2). A significantly higher mortality rate from the initial visit was observed in the period before 1982 ($P<0.01$), mainly due to difficulties in diagnosis (82% of patients who died before resection were seen before 1982). Postsurgical mortality was found to be similar between the two periods ($P=0.10$). Since 1982, the presence of a single ventricle was found to be the main factor associated with mortality ($P<0.01$).

Cardiac structure measures

Analyses of repeated echocardiography before and after membrane resection have shown that in biventricular patients, right ventricular systolic pressure (Figure 3), pulmonary valve size (Figure 4A), right and left ventricular end-diastolic dimension (Figure 5), and right and left pulmonary artery size show a trend toward normal after diaphragm resection without reaching statistical significance. Aortic valve size and tricuspid valve size showed a trend toward a moderate reduction of the z-score over time (Figures 4A and 4B), again, without reaching statistical significance. If compared with preoperative echocardiographic measurements, only mitral valve size did not change significantly for postoperative measurements (Figure 4B).

DISCUSSION

To our knowledge, the present study has investigated the largest group of patients with the diagnosis of CTS and subsequent outcomes in a single institution. Over the 50 years of the study period, patients presented with a variety of symptoms, sometimes associated with other cardiac defects. Symptoms at presentation depend mainly on the actual size of the perforation of the diaphragm, the hemodynamics and the atrial septal level (8).

Several classifications of CTS have been proposed, emphasizing anatomical and hemodynamic characteristics. Loeffler (12) distinguished three groups. The first group is characterized by the presence of two compartments in the left atrium and the absence of a communication between the upper space (receiving the pulmonary veins) and the lower chamber (which carries the appendage and opens into the left ventricle). This group of patients carries the highest risk of mortality and patients usually die during infancy. The second group involves patients with one or more small openings within the diaphragm. The third group includes patients in whom the accessory chamber communicates greatly with the left atrium and patients may live their

Figure 4) Change over time in z-score (size) for pulmonary and aortic valves (A), and mitral and tricuspid valves (B) measured by echocardiography after surgery. The solid lines represent the estimates of change in z-scores over time and the dashed lines represent the 95% CIs. P values are the estimates of the statistical significance of the observed time trends

entire life without intervention. Other classifications place more emphasis on the opening in the septum, separating patients into the following groups: those with no opening, a small opening or a wide opening (13). Lucas et al (14) combined elements of several classification systems, including connection of the pulmonary veins.

In a very small number of patients, the correct diagnosis of cor triatriatum sinistrum was made before referral, and may have been difficult even with the availability of the appropriate imaging tools (angiography and, later, echocardiography).

There was also a noteworthy shift in the mode of diagnosis over the study period. In the early years, cardiac catheterization, surgery or autopsy provided the correct diagnosis. However, with the implementation of ultrasound, a diagnosis by M-mode echocardiography was possible (15). Since the late 1980s, cross-sectional echocardiography decreased the number of delayed diagnoses and also became a more accurate method to delineate the defect (16). Later, transesophageal echocardiography further improved anatomical and intraoperative assessment of left heart structures (17,18). Echocardiographic findings consist of a usually thin intra-atrial membrane that may move throughout the cardiac cycle. CTS is usually visualized in multiple planes and can be differentiated from a supravulvar ring (16). Doppler echocardiography may also help to estimate the gradient across the left atrial diaphragm.

Of interest, patients who were discharged without intervention were all diagnosed after 1982, which strongly suggests that before that time, only the most severe cases were identified and patients with mild functional CTS probably went undiagnosed.

Figure 5) Changes over time in right and left ventricular end-diastolic dimensions after surgery in patients with cor triatriatum sinistrum. The solid lines represent the estimates of change in z-scores over time and the dashed lines represent the 95% CIs. P values are the estimates of the statistical significance of the observed time trends

The complete spectrum of congenital heart disease is not represented in our patient cohort; septation defects and pulmonary vein pathology dominate, whereas, for example, transposition of the great arteries, tetralogy of Fallot or truncus arteriosus were not diagnosed.

Primary treatment for CTS is surgery when needed. There are only two reports in the literature in which a catheter-based intervention was performed (19,20). Death after surgical repair of CTS is uncommon and is usually related to accompanying complex congenital heart disease. Mortality rates in other series that included both pediatric and adult patients, covering 1959 to 1992, ranged from 8% to 29% (4,5,7,8,10,11,21), and decreased to 4% in a more recent outcome analysis (9).

The overall mortality in our series was 19 patients (23%), with a higher proportion in the earlier time period ($n=12$), which equals 60% of the patients diagnosed between 1956 and 1981, compared with seven deaths (11% of all patients diagnosed) between 1982 and 2006. Fifteen of the 19 patients had other important structural congenital heart disease, including partial and total anomalous venous return, transposition of the great arteries with pulmonary stenosis and ventricular septal defect, single-ventricle physiology, coarctation of the aorta and pulmonary vein stenosis. Death before surgery was related to the complexity of the lesion and limited surgical options in the early cohort, or pulmonary infections. Two deaths were related to the cardiac catheterization procedure and unrelated to the CTS, one patient died following abdominal surgery, and for two patients, care was withdrawn due to hematological-oncological diseases.

There was almost no change in the mitral valve z-score over time, whereas the z-score for the pulmonary valve decreased to some extent. The aortic and tricuspid valve changes are most likely related to changes in flow and hemodynamics. This is further underlined by the drop in right ventricular pressure, which seems to plateau after five years.

CONCLUSION

The results strongly suggest that resection of the diaphragm allowed patients who survived the operation to regain near-normal cardiac dimensions. Despite generalized improvement of surgical technique and perioperative care, the frequency of complications was almost unchanged for both observation periods. This may be related to improved methods for tracking complications in the more recent era.

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