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ORIGINAL RESEARCH

RHYTHM DISORDERS AND ELECTROPHYSIOLOGY

Atrial and Ventricular Arrhythmia in Adults With Shone Complex

A Single-Center Cohort Study

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ABSTRACT

BACKGROUND Multilevel obstruction in left ventricular inflow and outflow predisposes to arrhythmias in Shone's complex (SC).

OBJECTIVES The purpose of this study was to study the prevalence and outcomes (heart failure [HF] hospitalization, cardiac transplant, death) of cardiac arrhythmias in adults with SC.

METHODS Adults with SC (defined as ≥2 lesions out of supramitral ring, parachute mitral valve, subvalvular/valvular aortic stenosis (AS), and aortic coarctation) seen at Mayo Clinic between January 1999 and March 2020 were identified and evaluated for the presence of sustained atrial fibrillation, atrial flutter, and ventricular arrhythmias (VA). Kaplan-Meier survival analysis was used to calculate the occurrence of these arrhythmias.

RESULTS Seventy-three patients with SC (mean age at first visit 33 ± 13 years) were identified. Most common anomalies were valvular AS (88%), coarctation (85%), parachute mitral valve (44%), subvalvular AS (44%), and supramitral ring (25%). Atrial arrhythmias were diagnosed in 24 patients (33%) at a mean age of 34.6 ± 12.7 years. Patients with atrial fibrillation and atrial flutter had higher number of surgeries, left atrial size, right ventricular systolic pressure, and HF hospitalizations. A rhythm control approach was used in majority of patients (75% on antiarrhythmic drugs and 50% underwent catheter ablation). Sustained VA occurred in 6 of 73 patients of whom 4 had an ejection fraction <40%. Death and cardiac transplantation occurred in 11 and 3 patients, respectively, during a median follow-up of 7.3 ± 6.0 years.

CONCLUSIONS In adults with SC, atrial arrhythmias occurred in one-third of patients, were associated with more HF hospitalizations, and frequently required rhythm control. Prevalence of sustained VA was 8% and implantable cardioverter-defibrillator implantation should be considered in those with reduced ejection fraction. (JACC Adv 2024;3:100715) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Shone's complex (SC) represents a constellation of congenital anomalies characterized by the presence of multilevel obstruction in the left-sided circulation. Originally described by Shone et al[1] in 1963, complete SC consists of 4 anomalies: supravalvular ring in the left atrium, muscular/membranous type of subaortic stenosis, parachute deformity of the mitral valve apparatus, and aortic

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ABBREVIATIONS AND ACRONYMS

AF = atrial fibrillation

AFL = atrial flutter

AV = atrioventricular

CIED = cardiac implanted electronic device

EF = ejection fraction

HF = heart failure

ICD = implantable cardioverter-defibrillator

LV = left ventricle

SC = Shone's complex

SVC = superior vena cava

TR = tricuspid regurgitation

VA = ventricular arrhythmias

VT = ventricular tachycardia

coarctation. The majority, however, have only 2 or 3 of these anomalies.

Advances in surgical management of congenital heart disease have resulted in more patients surviving to adulthood. Although the presence of left-sided obstructive lesions, need for multiple surgeries, and predisposition to heart failure (HF) portend a high burden of atrial and ventricular arrhythmias, this has not been well characterized in adults with SC. Accordingly, we sought to study the incidence and outcomes of atrial and ventricular arrhythmias in adults with SC treated at the Mayo Clinic.

METHODS

The Mayo Clinic electronic medical records were searched for patients ≥18 years of age who received the diagnosis of SC between January 1999 and March 2020. The diagnosis was based on the presence of an anatomically abnormal mitral valve and at least 1 additional left-sided obstructive lesion (fixed subaortic stenosis, valvular aortic stenosis, and/or aortic coarctation). Associated congenital heart diseases were also identified. The protocol was reviewed and approved by the Mayo Clinic Institutional Review Board. All patients consented to the use of their medical records for research purposes. Patients or the public were not involved in the design, conduct, reporting, or dissemination plans of our research.

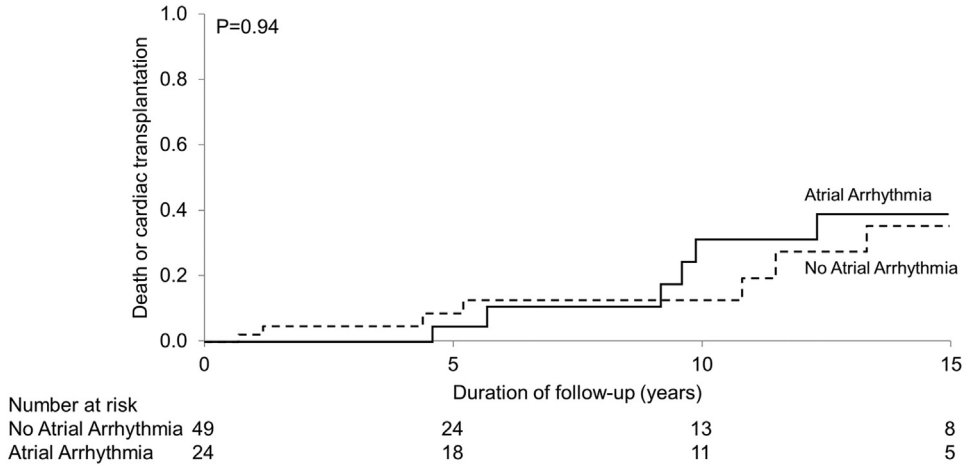
A retrospective review of Mayo Clinic records was conducted to identify clinical, surgical, echocardiographic and laboratory parameters. The first clinic encounter during the study period was defined as the index visit. Surgical reports and medical records prior to first visit at Mayo Clinic were also reviewed when available. The occurrence of sustained atrial arrhythmia lasting over 30 seconds at the time of first visit and during follow-up was identified by review of electrocardiograms, Holter monitors, cardiac implanted electronic device (CIED) interrogation, and review of clinical notes. Atrial fibrillation (AF), atrial flutter (AFL), and atrial tachycardia were distinguished for this study. Atrial arrhythmias occurring within 3 months of cardiac surgery were excluded. Sustained ventricular arrhythmias (VAs) including ventricular tachycardia (VT) and ventricular fibrillation lasting ≥30 seconds or causing hemodynamic instability (including syncope) were identified. Sudden cardiac death was defined as unexpected death occurring within 1 hour of the onset of symptoms or an unexpected unwitnessed death in a patient known to have been well at 1 time within the previous 24 hours.[2] Implantation of CIED including pacemakers and defibrillators was recorded. The diagnosis of acute HF was made by per chart review based upon the impression of the physician caring for the patient at that time. If these symptoms were accompanied by the sudden onset of or increase in the burden of AF with no other precipitating cause present, the HF was attributed to AF. Occurrence of death or cardiac transplantation was recorded, and participants were censored at that point. Survival status was ascertained using the Mayo clinic registration database and Accurint,[3] an institutionally approved fee-based Internet research location service that complies with the federal regulations concerning HIPAA (Health Insurance Portability and Accountability Act).

STATISTICAL ANALYSIS. Continuous variables are summarized using mean ± SD and compared using the Kruskal-Wallis test. Categorical variables are presented as number (percentage) and compared using the chi-square test. The cumulative incidence of the clinical outcomes of mortality and transplant, as well as hospitalization during follow-up, was estimated using the Kaplan-Meier method and log-rank tests. A 2-tailed P value ≤0.05 was considered statistically significant. All analyses were performed using SAS version 9.4, JMP version 16.1, and R version 4.0.3 or higher (R Foundation for Statistical Computing).

RESULTS

PATIENT CHARACTERISTICS. A total of 73 patients with SC were identified, with a mean age at the time of first visit of 33 ± 13 years and 59% females. The most common outflow anomalies were valvular aortic stenosis (n = 64, 88%), coarctation of aorta (n = 62, 85%), and subvalvular aortic stenosis (n = 32, 44%). The most common mitral valve anomalies included parachute mitral valve in 32 (44%) patients, supravalvular mitral membrane in 18 (25%) patients, dysmorphic or abnormal mitral valve leaflets leading to valvular stenosis in 23 (32%). Other mitral valve abnormalities included shortened chordae and abnormal subvalvular apparatus (n = 4), double orifice mitral valve (n = 3), mitral arcade (n = 1), cleft mitral valve (n = 1), and mitral valve prolapse (n = 2). The mitral valve abnormality was unknown in 4 patients who had undergone previous mitral valve replacement. An accompanying ventricular septal

FIGURE 1 Time to Occurrence of Death or Cardiac Transplantation in Shone's Complex Stratified by Presence of Atrial Arrhythmias



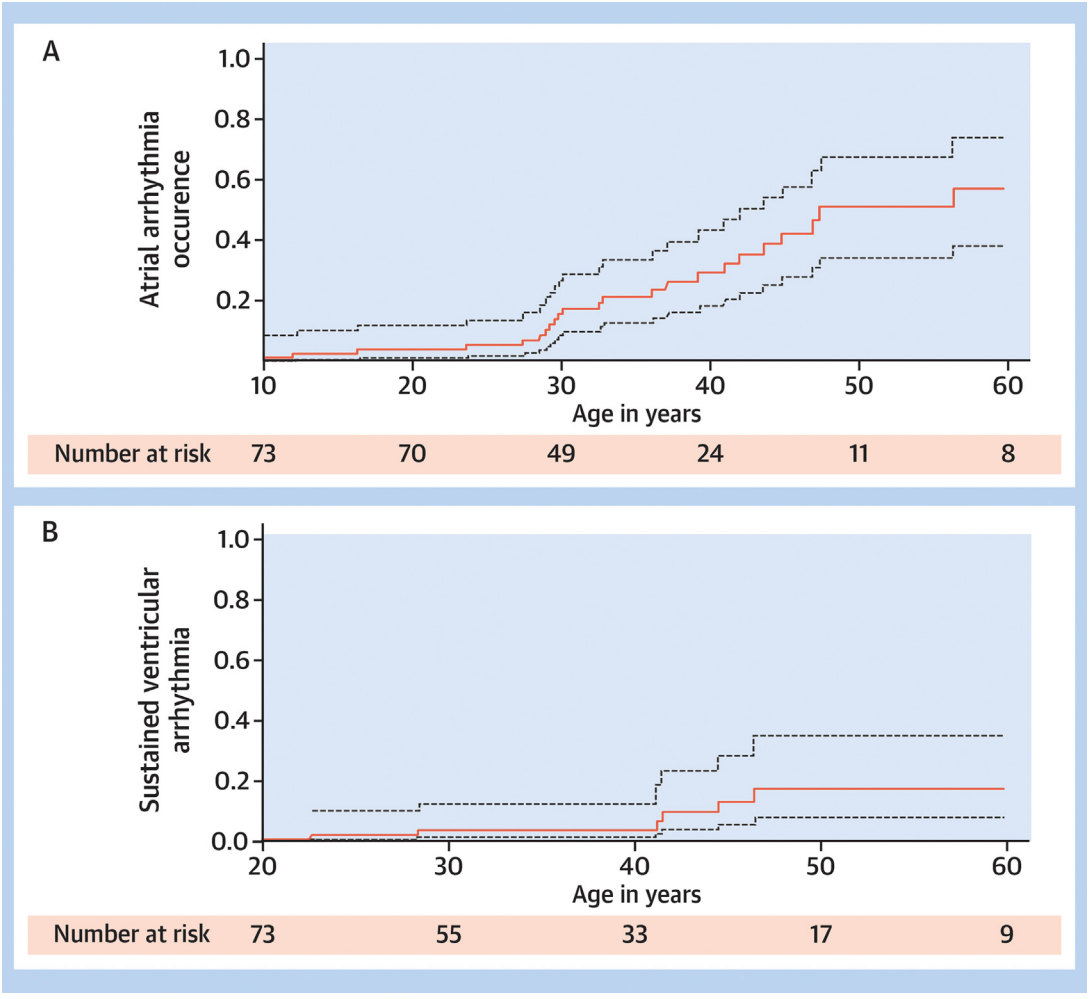
defect was present in 19 (26%) patients. Other associated congenital anomalies included a persistent left-sided superior vena cava (SVC) (n = 15), ruptured sinus of Valsalva (n = 1), anomalous coronary artery anatomy (n = 6), hypoplastic aortic arch (n = 4), interrupted aortic arch (n = 2), and partial anomalous pulmonary venous return (n = 3).

ATRIAL ARRHYTHMIAS. At the time of first presentation, 11 (15%) patients had a history of atrial arrhythmias. During the median follow-up of 7.3 ± 6.0 years, an additional 13 (18%) patients were diagnosed with one or more atrial arrhythmias, resulting in a prevalence of 24 (33%) at the last follow-up. Isolated AF was diagnosed in 7 (29%), isolated AFL in 4 (16.6%), combined atrial fibrillation and flutter in 11 (46%), and paroxysmal supraventricular tachycardia in 2 (8%). The mean age of diagnosis of atrial arrhythmia was 34.6 ± 12.7 years and there was no difference in age between those with (36.3 ± 11.8 years) and those without (32.4 ± 13.3 years) atrial arrhythmias ($P = 0.082$). The time to the occurrence of atrial arrhythmias in these patients is depicted in **Central Illustration A**. Atrial arrhythmia was paroxysmal in 20 (83%), persistent in 3 (13%), and permanent in 1 (4%) patient. The main symptoms of atrial arrhythmia included palpitations (n = 14, 58%) and dyspnea (n = 12, 50%). Chest pain, fatigue, and hemodynamic instability were noted in 2 patients each. No symptoms were reported in 5 patients. Patients with atrial arrhythmias were more likely to be hospitalized for HF compared to those without atrial arrhythmias (46% vs 22%; $P = 0.040$).

Baseline characteristics of the whole cohort and stratified by the presence or absence of atrial arrhythmias are presented in **Table 1**. Patients with atrial arrhythmias were more likely to have unrepaired mitral valve anomaly, greater number of cardiac surgeries, larger left atrial size, higher right ventricular systolic pressure, and higher likelihood of ≥ moderate tricuspid regurgitation (TR) compared to those without atrial arrhythmias.

Rhythm control with at least one antiarrhythmic drug was pursued in the majority of patients (n = 18/24, 75%), with 10 patients requiring more than 1 antiarrhythmic medication. Antiarrhythmic drugs used included amiodarone (n = 16), dofetilide (n = 4), sotalol (n = 4), dronedarone (n = 1), quinidine (n = 1), flecainide (n = 1), and propafenone (n = 3). One or more cardioversions were performed in 20 of 24 (83.3%) patients. In addition, 10 patients underwent a total of 14 percutaneous catheter ablation procedures, involving pulmonary vein isolation (n = 5), cavotricuspid isthmus ablation (n = 4), and atrioventricular (AV) node ablation (n = 3). Atypical flutter was ablated in some, including right atriotomy scar dependent flutter (n = 1), left atrial roof (n = 2), and mitral isthmus (n = 2) dependent flutter. AV nodal reentrant tachycardia (n = 2) and focal atrial tachycardia (n = 2) were also ablated in a minority of patients. Surgical MAZE procedure was performed in 3 patients. Recurrence of atrial arrhythmia, predominantly AFL and fibrillation, occurred in 8/10 patients who underwent catheter or surgical ablation for rhythm control at 1 year following ablation.

CENTRAL ILLUSTRATION Atrial and Ventricular Arrhythmias in Shone's Complex



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Time to occurrence of (A) atrial arrhythmia and (B) sustained ventricular arrhythmia in Shone's complex.

Anticoagulation for stroke prevention was prescribed to 21 of 24 patients. Warfarin was most commonly used (n = 18) and a direct oral anticoagulant was prescribed in 3 patients. Incidence of stroke was not significantly different between those with (n = 2) and without (n = 4) atrial arrhythmias ($P = 0.982$).

VENTRICULAR ARRHYTHMIA. A total of 6 of 73 (8%) patients in our cohort were noted to have sustained VAs. Seven patients had an implantable cardioverter-defibrillator (ICD) implanted: 3 for secondary prevention and 4 for primary prevention of sudden cardiac death in the setting of left ventricular systolic dysfunction and ejection fraction (EF) <35% (Central Illustration A). The time to first sustained VT or ICD therapy for VT is presented in Central Illustration B.

Of the 73 patients, 5 patients had LV EF<35%, of whom 4 underwent primary prevention ICD implantation. During follow-up, 3 of the 4 patients with a primary prevention ICD received at least one ICD shock for polymorphic VT and all 3 were subsequently treated with amiodarone. One patient with primary

TABLE 1 Baseline Characteristics of the Entire Cohort and Stratified by the Occurrence of Atrial Arrhythmias

		Arrhythmia Status		
	Whole Cohort (N = 73)	No Atrial Arrhythmia (n = 49)	Atrial Arrhythmia (n = 24)	*P* Value
Age at first visit (y)	33 ± 13	32 ± 13	36 ± 11	0.080
Female	43 (58.9)	29 (59.2)	14 (58.3)	0.942
Left-sided obstructive lesions				
Coarctation of the aorta	62 (84.9)	42 (85.7)	20 (83.3)	0.781
Valvular aortic stenosis	64 (87.7)	44 (89.8)	20 (83.4)	0.520
Subvalvular aortic stenosis	33 (43.8)	24 (49)	8 (33.4)	0.212
Mitral valve morphologies				
Parachute	32 (43.8)	23 (46.9)	9 (37.5)	0.450
Supramitral ring	18 (24.6)	12 (24.4)	6 (25.0)	0.953
Dysmorphic	13 (17.8)	7 (14.2)	6 (25.0)	0.260
Valve stenosis	10 (13.6)	10 (13.6)	0	
Subvalvular abnormality	4 (5.4)	2 (4.0)	2 (8.3)	0.454
Double orifice	3 (4.1)	3 (4.1)	0	
Valve prolapse	2 (2.6)	2 (2.6)	0	
Calcified valve	2 (2.6)	1 (2.0)	1 (4.1)	0.60
Cleft valve	1 (1.3)	1 (1.3)	0	
Arcade	1 (1.3)	1 (1.3)	0	
Unknown underlying anatomy	4 (5.4)	0	4 (5.4)	
History of cardiac surgery				
Number of cardiac surgeries	3 (2-4)	2 (1-3)	3 (2.5-4)	0.020
Surgery for mitral valve anomaly	19 (26.0)	6 (12.2)	13 (54.2)	<0.001
Age at first coarctation of aorta surgery (y)	2 (0-6)	1 (0-6)	2 (0-9)	0.372
Age at first surgery for valvular aortic stenosis (y)	16 (7-33)	18 (8-34)	11 (4-25)	0.261
Age at first subvalvular aortic stenosis surgery (y)	6 (3-9)	6 (3-8)	7 (3-9)	0.911
Age at first mitral surgery (y)	17 (6-31)	21 (7-33)	12 (4-30)	0.322
Comorbidities				
Hypertension	18 (24.7)	15 (30.6)	3 (12.5)	0.091
History of stroke	6 (8.2)	4 (8.2)	2 (8.3)	0.980

Values are mean ± SD, n (%), or median (IQR).

prevention ICD also received an inappropriate shock for sinus tachycardia. All patients with low EF and ICD implantation also had a history of surgery for either aortic coarctation, aortic valve replacement, or mitral valve replacement. In addition, 1 patient had a history of circumflex artery injury during mitral valve surgery due to anomalous course of the artery near the supramitral ring.

Sustained monomorphic VT associated with syncope or palpitations occurred in 3 patients without a pre-existing ICD leading to implantation of a secondary prevention ICD. One patient with EF <40% and a history of surgical coronary artery injury was treated with sotalol and mexiletine without subsequent VT recurrence. Another patient with EF of 60%, repaired aortic coarctation, ruptured sinus of Valsalva, and mitral valve replacement had wide complex tachycardia. Programmed ventricular stimulation did not induce VT, but a secondary prevention ICD was placed without any subsequent ICD therapies or antiarrhythmic drug therapy. Finally, a third patient with EF 70%, prior repair of aortic coarctation and aortic valve replacement had ICD implanted following occurrence of monomorphic VT. Although there were no known ICD therapies, sudden cardiac death occurred, and cause of death could not be ascertained in the absence of ICD interrogation or autopsy.

CIED IMPLANTATION. The proportion of patients out of the entire cohort who received a CIED (stratified by atrial arrhythmia) is presented in Table 2. In addition to the 7 patients undergoing ICD implantation, 15 patients (22%) had a pacemaker device implanted. Indications included: 1) AV block (n = 11) following cardiac surgery; 2) sinus node dysfunction (n = 2); and 3) AV node ablation for atrial arrhythmia (n = 2). A left-sided SVC was present in 15 (20.5%) patients in the cohort. Six patients with left SVC underwent device implantation, 5 through the right subclavian vein. One patient initially had a CIED implanted through the left SVC, but this was later revised to the right subclavian vein due to lead dislodgment.

Following CIED implantation, short-term complications (<3 months) included pocket hematoma requiring evacuation in 2 patients and lead dislodgment requiring revision in 2 patients. Long-term complications included one or more lead revisions due to lead malfunction in 4 patients, pocket revision due to pain in 1 patient, device infection leading to extraction and reimplantation in 1 patient and severe tricuspid valve regurgitation due to impingement of CIED lead on valve leaflet requiring tricuspid valve repair or replacement with exteriorization of the lead to the valve in 1 patient.

DEATH AND CARDIAC TRANSPLANTATION. **Figure 1** demonstrates the death and cardiac transplantation outcomes for the cohort across their follow-up period, stratified by atrial arrhythmias. A total of 3 patients underwent a cardiac transplant and 11 died by the end of the follow-up period. Of the latter group, only 2 were known to have died due to known cardiac causes. The first patient suffered a myocardial infarction and the second had a sudden cardiac death, having had previous episodes of monomorphic VT and prior ICD implantation. **Table 2** summarizes the outcomes for the entire cohort based on the presence/absence of atrial arrhythmia during their follow-up period.

DISCUSSION

In the largest study to systematically analyze atrial and ventricular arrhythmias in adults with SC to date, we report: 1) atrial arrhythmias, predominantly AFL and fibrillation, in one-third of patients with mean age of presentation of 35 years; 2) multiple cardiac surgeries, unrepaired mitral valve anomaly, pulmonary hypertension, and large left atrial size were associated with atrial arrhythmias; 3) the majority of patients with atrial arrhythmias were symptomatic and received multiple interventions to restore rhythm including anti-arrhythmic drugs, cardioversion, and ablation; 4) LV EF <35% was more common in those with sustained VAs. supporting implantation of ICD for primary prevention of sudden cardiac death in these patients; and 5) CIED implantation was necessary in 22 patients (15 with pacemaker and 7 with ICD).

Patients with SC have multilevel obstruction in the left-sided circulation frequently requiring cardiac surgery for repair of aortic coarctation, subaortic stenosis, and aortic or mitral valve replacement. The severity and management of mitral anomaly have prognostic significance in SC and the severity of mitral stenosis correlates with increased mortality cardiac surgery.[4,5] In our cohort, surgery for LV outflow obstruction was commonly performed in the first decade of life, but mitral valve interventions were usually delayed until the second decade or later. Thus, the high prevalence of atrial arrhythmias in one-third of this young cohort is presumably secondary to persistent multilevel obstruction, particularly mitral inflow obstruction. Moreover, AF or AFL occurred at a young age, with the mean age being 35 years. Aslam et al have reported AF and AFL in 14% and paroxysmal supraventricular tachycardia in 11% of patients in a cohort of 28 SC patients with a mean age of 25 years. The lower prevalence of AF and AFL is likely related to younger age of their cohort compared to the current study.[6] The high prevalence of atrial arrhythmia in SC parallels that reported in some complex congenital heart diseases. For instance, in a cohort of adult patients with Fontan circulation with a mean age of 29 years, Quinton et al[7] reported a 42% prevalence of atrial arrhythmias.

The potential pathogenic mechanisms of atrial arrhythmias in SC include: 1) chronic left atrial volume and pressure overload leading to adverse electrical and structural remodeling with left atrial enlargement; 2) development of pulmonary hypertension leading to elevated right-sided pressure, right ventricular dysfunction, and TR; and 3) presence of atriotomy scar following cardiac surgery predisposing to reentrant flutters. This is borne out in our study by the more frequent presence of unrepaired mitral anomaly, mitral valve surgery, multiple prior cardiac surgeries, higher right ventricular systolic pressure, and TR in patients with atrial arrhythmias. Beyond these traditional mechanisms for AF in mitral stenosis, the presence of multilevel outflow obstruction in SC may also lead to LV diastolic dysfunction, which further compromises left atrial emptying, thus contributing to left atrial volume and pressure overload.[8] The presence of mitral stenosis and LV diastolic dysfunction could also explain the high prevalence of symptoms during atrial arrhythmias. Tachycardia and irregular R-R intervals further compromise LV diastolic filling and increase pulmonary pressure leading to dyspnea, fatigue, and HF, as was frequently noted in our study. Atrial arrhythmias were associated with higher likelihood of greater than moderate TR. While TR may contribute to the development of atrial arrhythmias, recent data also suggest that atrial arrhythmias may contribute to worsening TR.[9,10]

Patients with SC having AF or AFL frequently experience symptoms and were more prone to develop HF than non-SC patients. The majority of patients in our study required treatment for rhythm control of AF and/or AFL. Antiarrhythmic drugs were

TABLE 2 Medication Use, Echocardiographic Parameters, Device Implantation, and Clinical Outcomes During Follow-Up of the Entire Cohort and Stratified by the Occurrence of Atrial Arrhythmias

		Arrhythmia Status		
	Whole Cohort (N = 73)	No Atrial Arrhythmia (n = 49)	Atrial Arrhythmia (n = 24)	*P* Value
Medications				
Beta-blocker	30 (41.1)	19 (38.8)	11 (45.8)	0.562
Calcium channel blocker	7 (9.6)	4 (8.2)	3 (12.5)	0.550
Angiotensin-converting enzyme inhibitor or angiotensin receptor blocker	18 (24.7)	8 (16.3)	10 (41.7)	0.018
Oral anticoagulation	43 (58.9)	22 (44.9)	21 (87.5)	<0.001
Echocardiogram				
LV ejection fraction (%)	60 ± 11	61 ± 9	56 ± 15	0.210
Right ventricular systolic pressure (mm Hg)	47.7 ± 15.5	44.3 ± 14.5	53.7 ± 15.8	0.008
≥Moderate tricuspid regurgitation	11 (15.1)	4 (8.2)	7 (29.2)	0.020
Left atrial volume index (mL/m^2)	40.4 ± 24.7	34.8 ± 18.2	52.7 ± 32.4	0.002
LV outflow tract mean gradient (mm Hg)	22.0 ± 19.2	24.6 ± 20.8	16.7 ± 14.3	0.172
Coarctation of aorta mean gradient (mm Hg)	14.3 ± 9.9	15.3 ± 11.2	12.3 ± 6.5	0.373
Cardiac device implantation				
Pacemaker implantation	15 (21.9)	8 (16.7)	8 (33.3)	0.060
Implantable cardioverter-defibrillator implantation	7 (9.5)	3 (6.1)	4 (16.6)	0.594
Outcomes				
Heart failure hospitalization	22 (30.1)	11 (22.4)	11 (45.8)	0.040
Death	11 (15.1)	5 (10.2)	6 (25.0)	0.100
Transplant	3 (4.1)	2 (4.1)	1 (4.2)	0.991

Values are n (%) or mean ± SD. The medications mentioned in the table were used during the period of our follow-up for the patients in our cohort. The echocardiographic data pertain to the procedure performed at the index visit.

LV = left ventricle.

used in most patients and catheter, or surgical ablation was also frequently performed. Recurrences were frequent following drug and ablation therapy, underscoring the difficulty in maintaining sinus rhythm in these patients. A small number of patients in our cohort required AV node ablation following placement of a pacemaker to alleviate symptoms. Optimal rhythm control strategies and the outcomes of catheter ablation need further study.

This is the first detailed report of the occurrence of VAs in SC, with sustained hemodynamically significant VT occurring in 8% of patients. Potential pathogenic mechanisms underlying VT may include LV pressure overload leading to myocardial fibrosis, LV systolic dysfunction, and rarely coronary artery injury during surgery as was noted in 2 patients in this cohort. LV systolic dysfunction with EF <40%, noted in 4 of 6 patients with documented VT, emerged as a potential, though plausible risk factor. Thus, the current recommendation[11] to consider primary prevention ICD implantation in patients with LV EF ≤35% appears to be applicable to SC patients also. It is notable that 75% of SC patients receiving a primary prevention ICD for low EF subsequently had ICD shock, an incidence higher than that reported in adults with primary prevention ICDs for common forms of ischemic or nonischemic cardiomyopathy. For example, 21% of individuals with an ICD in the Sudden Cardiac Death in Heart Failure Trial received an appropriate shock.[12] However, the current cohort is too small to draw firm conclusions regarding the risk of sudden death and requires further investigation in a large multicenter study.

STUDY LIMITATIONS. Our study has limitations inherent to retrospective and observational studies. This includes the inability to comment on relative efficacy of different treatment modalities for atrial and ventricular arrhythmias including ablation and ICD implantation. Although this is the largest published cohort of SC, the small sample size of this rare condition limits our ability to identify definitive risk factors for the development of atrial and ventricular arrhythmias using multivariable analyses. Although patients followed at our center undergo periodic routine testing including Holter, electrocardiogram, and echocardiogram, the duration and techniques used for follow-up were not predefined. It is also possible that some episodes of atrial arrhythmias that were asymptomatic may have been missed.

CONCLUSIONS

Atrial arrhythmias were observed in one-third of patients with SC and were associated with unrepaired mitral anomaly, pulmonary hypertension, and multiple cardiac surgeries. Prompt recognition and treatment of AF or AFL is necessary to alleviate associated symptoms. SC patients with reduced left ventricular ejection fraction are at high risk of sustained VT and should be considered for ICD implantation. Future prospective multicenter studies are required to further characterize the risk factors for arrhythmias and their response to treatment.

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PERSPECTIVES

COMPETENCY IN MEDICAL KNOWLEDGE: It is well-known that the occurrence of multilevel obstruction in the left ventricular inflow and outflow may predispose to atrial and ventricular arrhythmias in SC. The presence of these arrhythmias is expected to greatly contribute to the morbidity and mortality of these patients.

TRANSLATIONAL OUTLOOK: In our study, atrial arrhythmias occurred in one-third of patients and were associated with more heart failure hospitalizations and rhythm control interventions. Sustained ventricular arrhythmias were observed predominantly in those with reduced left ventricular ejection fraction and these patients should be considered for implantation of cardioverter defibrillator. Therefore, an increased awareness of these arrhythmic complications is essential to ensure timely diagnosis and referral to specialized centers for appropriate management.

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