Received: 2 January 2019 | Revised: 8 June 2019 | Accepted: 16 July 2019
DOI: 10.1111/chd.12833

ORIGINAL ARTICLE

Congenital Heart Disease WILEY

# Improvement in ventricular function with rhythm control of atrial arrhythmias may delay the need for atrioventricular valve surgery in adults with congenital heart disease

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**Funding information**
Supported by the Pennsylvania Steel Company EP Research Fund.

## Abstract

**Objective:** Atrial arrhythmias and atrioventricular valve regurgitation (AVVR) are common causes of morbidity among adults with congenital heart disease (ACHD). The impact of rhythm control on AVVR in this population is unknown. We sought to determine whether a rhythm control strategy is associated with greater freedom from AV valve surgery than a rate control strategy.

**Design:** Patients evaluated by both ACHD and electrophysiology specialists at a single academic center were screened for atrial arrhythmias and at least moderate-severe AVVR. Clinical and electrographic data were abstracted. All echocardiograms were interpreted by a single echocardiographer blinded to treatment strategy. Patients were followed until AV valve surgery, heart transplantation, death, or last clinical follow-up.

**Results:** Rhythm control was attempted in 9 of 24 identified patients. Among these nine patients, arrhythmias were eliminated in three and reduced from persistent to paroxysmal in another three. In the rhythm control group, mean left ventricular ejection fraction improved from 54.4 ± 12.4% to 60.0 ± 11.5% ($P$ = .02) and mean right ventricular systolic function increased nearly one grade ($P$ = .02). AVVR did not decrease significantly. No significant change in left or right ventricular systolic function, or AVVR was observed among the 15 patients treated with rate control. Four-year survival free of AV valve operation and heart transplant was 88% in the rhythm control group and 31% in the rate control group ($P$ = .04).

**Conclusions:** In ACHD patients with atrial arrhythmias and at least moderate-severe AVVR, a rhythm control strategy was associated with improved biventricular systolic function. This improvement in ventricular function and symptoms may allow valve surgery to be deferred.

KEYWORDS
adult congenital heart disease, atrial arrhythmias, intra-atrial reentrant tachycardia, rhythm control, supraventricular tachycardia, valve regurgitation

**Abbreviations:** ACHD, adult congenital heart disease; AV, atrioventricular; AVVR, atrioventricular valve regurgitation; IART, intra-atrial reentrant tachycardia.

  

# 1 | INTRODUCTION

Atrial arrhythmias affect 15% of adults with congenital heart disease (ACHD), with a lifetime cumulative risk >50% for patients with severe cardiac lesions.[1] As more patients with congenital heart disease survive later into adulthood, atrial arrhythmias are becoming an increasingly important cause of morbidity and mortality.[2-7] The predisposition to atrial arrhythmias in ACHD is multifactorial, including postsurgical scarring and fibrosis, long-standing pressure and volume overload, and intrinsic conduction disease. These make rhythm control challenging, whether by antiarrhythmic drugs[8-10] or catheter ablation.[11-14]

Atrial arrhythmias frequently coexist with atrioventricular valve regurgitation (AVVR), both of which are associated with increased mortality in ACHD.[15] Classically, AVVR has been thought to drive adverse atrial remodeling thereby promoting arrhythmias; however, fixing AVVR does not always eliminate atrial arrhythmias.[16] Recent studies on "atrial functional AVVR," and its reversibility with rhythm control, suggest a more complex relationship between atrial arrhythmias and AVVR.[17,18] No studies to date have examined the impact of rhythm control of atrial arrhythmias on AVVR in the ACHD population.

We identified all patients with atrial arrhythmias and at least moderate-severe AVVR treated at a large ACHD center. We hypothesized that rhythm control, through either catheter ablation or antiarrhythmic drugs, would be associated with greater freedom from atrioventricular (AV) valve surgery.

# 2 | METHODS

## 2.1 | Study population

All patients evaluated by both an adult congenital cardiologist and cardiac electrophysiologist at the Hospital of the University of Pennsylvania between January 2004 and January 2017 were identified using the electronic medical record. These patients were then further screened for the presence of atrial arrhythmias and at least moderate-severe AVVR. Patients treated with catheter ablation or a class I or class III antiarrhythmic drug were included in the rhythm control group. All other patients were included in the rate control group. The study was approved by the University of Pennsylvania institutional review board.

## 2.2 | Classification of atrial arrhythmias

Atrial arrhythmias were classified as intra-atrial reentrant tachycardia (IART), atrial fibrillation, atrioventricular nodal reentry tachycardia, or focal atrial tachycardia by review of electrocardiograms, ambulatory monitors, and ablation reports. Arrhythmias were considered paroxysmal if self-terminating in <7 days and persistent if lasting ≥7 days or requiring direct current cardioversion. Patients underwent ambulatory monitoring at regular intervals, according to the practice of the treating clinician. Arrhythmia recurrence was defined as electrographically documented atrial arrhythmia lasting >30 seconds. Reduction in arrhythmia burden was defined as either no recurrence or decrease in arrhythmia pattern from persistent to paroxysmal.

## 2.3 | Echocardiographic measurements

All echocardiograms were performed by a sonographer with expertise in ACHD and reinterpreted for this research study by a single echocardiographer (Y.K.), blinded to the treatment group. Atrial size was characterized qualitatively for all patients, excluding those with intra-atrial baffles. Standard techniques were used to quantify right and left ventricular size and function, as well as valvular function.[19] Particular attention was devoted to changes in AVVR and ventricular systolic function over time. Ventricles were primarily analyzed according to their morphology (right or left). In secondary analyses, ventricles were analyzed according to their position (subsystemic or subpulmonic).

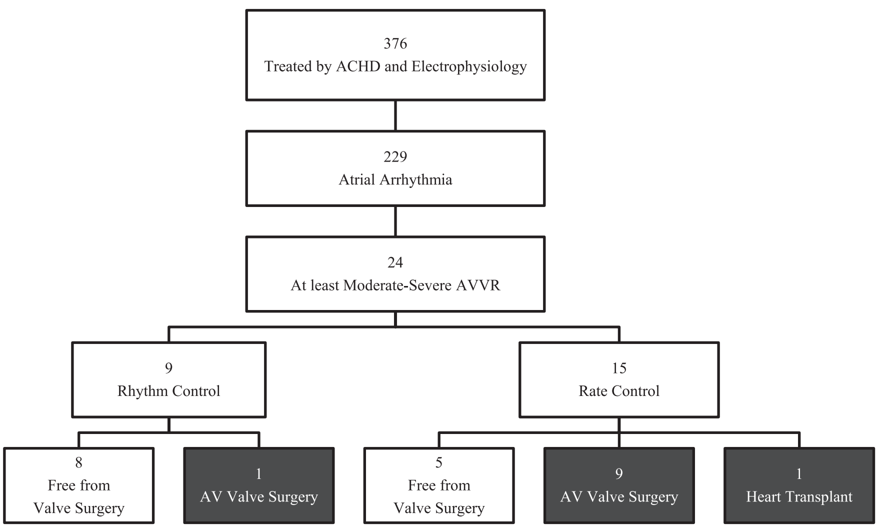


**FIGURE 1** Patient flow and outcomes. Patients with adult congenital heart disease (ACHD), atrial arrhythmias, and at least moderate-severe atrioventricular valve regurgitation (AVVR) were identified and then followed for AV valve surgery and heart transplant

**TABLE 1** Clinical characteristics of rhythm control and rate control cohorts

| Characteristic | Rhythm control (*n* = 9) | Rate control (*n* = 15) | *P* value |
|---|---|---|---|
| Male sex | 3 (33.3) | 7 (46.7) | .7 |
| Age (yr) | 47.0 ± 11.8 | 46.6 ± 15.0 | .9 |
| Congenital heart disease type | | | .5 |
| Ebstein's anomaly | 2 (22.2) | 0 | |
| Tetralogy of fallot | 2 (22.2) | 4 (26.7) | |
| D-TGA | 1 (11.1) | 3 (20.0) | |
| AV canal defect | 1 (11.1) | 2 (13.3) | |
| Atrial septal defect | 1 (11.1) | 1 (6.7) | |
| Ventricular septal defect | 2 (22.2) | 2 (13.3) | |
| Other | 0 | 3 (20.0) | |
| Congenital heart disease class | | | .8 |
| Simple | 1 (11.1) | 3 (20.0) | |
| Moderate | 6 (66.7) | 8 (53.3) | |
| Complex | 2 (22.2) | 4 (26.7) | |
| Systemic right ventricle | 1 (11.1) | 4 (26.7) | .6 |
| Prior operations | | | |
| Number of prior operations | 1.6 ± 0.7 | 1.8 ± 1.2 | .5 |
| AV valve operation | 1 (11.1) | 4 (26.7) | .6 |
| Cox maze | 1 (11.1) | 1 (6.7) | .9 |
| Years since last operation | 27.7 ± 14.3 | 18.2 ± 11.7 | .1 |
| Implantable cardiac devices | 2 (22.2) | 9 (60.0) | .1 |
| Permanent pacemaker | 1 (11.1) | 7 (46.7) | .2 |
| ICD | 1 (11.1) | 2 (13.3) | .9 |
| Comorbidities | | | |
| Any comorbidity | 4 (44.4) | 8 (53.3) | .9 |
| Chromosomal abnormalities | 1 (11.1) | 0 | .4 |
| Chronic kidney disease | 0 | 1 (6.7) | .9 |
| Coronary artery disease | 1 (11.1) | 0 | .4 |
| Diabetes mellitus | 0 | 2 (13.3) | .5 |
| Hypertension | 2 (22.2) | 2 (13.3) | .6 |
| Obesity | 1 (11.1) | 3 (20.0) | .9 |
| Pulmonary disease | 0 | 2 (13.3) | .5 |
| Thyroid disease | 0 | 3 (20.0) | .3 |
| QTc duration (ms) | 462 ± 29.8 | 470.4 ± 48.2 | .6 |

*Note:* Values are *n* (%) or mean ± standard deviation. Lesions in other congenital heart disease category include congenitally corrected transposition of the great arteries, anomalous pulmonary venous return, and pulmonary stenosis.
Abbreviations: D-TGA, dextro-transposition of the great arteries; ICD, implantable cardioverter-defibrillator.

## 2.4 | Clinical follow-up

Baseline was defined as the date of the first attempt at rhythm control or the first echocardiogram demonstrating at least moderate-severe AVVR in the rate control group. Demographic and clinical characteristics were extracted from the electronic medical record. Patients were followed longitudinally until AV valve surgery, cardiac transplantation, death, or last clinical follow-up, whichever occurred first.

## 2.5 | Statistical analysis

Continuous data are summarized as mean ± standard deviation or median (range) and categorical variables are presented as frequencies with percentages. Continuous variables were compared using the Student *t* test and dichotomous variables using Fisher's exact test. Kaplan-Meier curves were constructed to illustrate survival free from valve surgery and heart transplantation. Survival was compared between the rhythm and the rate control groups using the

**TABLE 2** Arrhythmia characteristics, interventions, and outcomes

| | Rhythm control (*n* = 9) | Rate control (*n* = 15) | *P* value |
|---|---|---|---|
| Arrhythmia type | | | |
| IART | 8 (88.9) | 6 (40.0) | .01 |
| Atrial fibrillation | 4 (44.4) | 10 (66.7) | .7 |
| AVNRT | 0 | 1 (6.7) | .9 |
| Focal atrial tachycardia | 2 (22.2) | 5 (33.3) | .9 |
| Baseline arrhythmia burden | | | .2 |
| Paroxysmal | 2 (22.2) | 7 (46.7) | |
| Persistent | 7 (77.8) | 8 (53.3) | |
| Beta blocker | 7 (77.8) | 9 (60.0) | .7 |
| Calcium channel blockers | 1 (11.1) | 1 (6.7) | .9 |
| Arrhythmia interventions | 2.6 ± 1.7 | x | |
| Antiarrhythmic drugs | 9 (100.0) | x | |
| Amiodarone | 4 (44.4) | x | |
| Dofetilide | 7 (77.8) | x | |
| Propafenone | 1 (11.1) | x | |
| Sotalol | 3 (33.3) | x | |
| Number of AADs | 1.78 ± 1.1 | x | |
| Catheter ablation | 4 (44.4) | x | |
| Number of catheter ablations | 1.5 ± 0.6 | x | |
| Heart rate control | | | |
| Average heart rate | 69.0 ± 7.5 | 74.1 ± 7.2 | .3 |
| Outcomes | | | |
| No recurrence | 3 (33.3) | 3 (20.0) | .6 |
| Paroxysmal recurrence | 4 (44.4) | 6 (40.0) | .9 |
| Persistent recurrence | 2 (22.2) | 6 (40.0) | .7 |
| Reduction in arrhythmia burden | 6 (66.7) | 3 (20.0) | .04 |

*Note:* Values are *n* (%) or mean ± standard deviation.
Abbreviations: AADs, antiarrhythmic drugs; AVNRT, atrioventricular nodal reentry tachycardia; IART, intra-atrial reentrant tachycardia.

log-rank test. Two-tailed *P* values < .05 were considered statistically significant. Analyses were performed using SPSS software (version 24, SPSS Inc., Chicago, IL).

## 3 | RESULTS

Of the 376 patients treated by both an adult congenital cardiologist and cardiac electrophysiologist over the study period, 229 had at least one documented atrial arrhythmia. Of these, 24 had at least moderate-severe AVVR. Nine were treated with rhythm control and the remaining 15 with rate control (Figure 1).

Subjects in the rhythm and rate control groups had similar baseline characteristics (Table 1). A wide spectrum of lesions was similarly distributed across both groups, with no difference in congenital heart disease complexity according to accepted classifications (*P* = .8).[20] There was no difference in the number of prior cardiac operations (*P* = .5). Medical comorbidities were uncommon. Baseline QTc was similar between the two groups (*P* = .6).

Patients in the rhythm control group were more likely to have IART (Table 2, *P* = .01). Arrhythmias were persistent in 77.8% of patients in the rhythm control group and 46.7% of patients in the rate control group (*P* = .2). Patients in the rhythm control group attempted a mean of 2.6 rhythm control interventions. All patients were treated with antiarrhythmic drugs, most commonly dofetilide (77.8%). Catheter ablation was performed in 44.4% of patients, with a mean of 1.5 ablations per patient. There were no complications among those undergoing ablation, including no periprocedural increases in valvular regurgitation. No difference was observed in strict freedom from atrial arrhythmia recurrence (*P* = .6). However, reduction in arrhythmia burden was more commonly achieved in the rhythm control group (66.7% vs 20.0% in the rate control group, *P* = .04).

Baseline left atrial size, left ventricular (LV) size, and ejection fraction were similar between both groups (Table 3). Similarly, right atrial size, right ventricular (RV) size, and function did not significantly differ at baseline. The tricuspid valve was the regurgitant valve in the majority of patients in both groups.

Follow-up echocardiograms were available for all patients, performed a mean of 2.2 years after baseline in the rhythm control group and 2.4 years after baseline in the rate control group (Table 4). Mean LV ejection fraction improved from 54.4 ± 12.4% to 60.0 ± 11.5% (*P* = .02) in the rhythm control group and decreased from 60.4 ± 11.1% to 58.9 ± 9.0% (*P* = .7) in the rate control group (Figure 2, panels A and B). Mean RV systolic function improved nearly one grade in the rhythm control group (*P* = .02) and decreased slightly in the rate control group (*P* = .4) (Figure 2, panels C and D). When analyzed according to position, there was significant improvement in subpulmonic ventricular systolic function (*P* = .05), and a nonsignificant trend toward improvement in subsystemic ventricular function in the rhythm control group (Supporting information Figure S1, panels A and C, *P* = .2). There were no improvements in systolic function in the rate control group (Supporting information Figure S1, panels B and D). The degree of AVVR did not significantly improve in either group.

One patient in the rhythm control group and nine patients in the rate control group underwent AV valve operation during the study period. An additional patient in the rate control group underwent heart transplant. Cumulative AV valve operation and transplant-free survival at 4-year follow-up was 88% in the rhythm control group and 31% in the rate control group (Figure 3, log-rank *P* = .04). Concurrent MAZE was performed in four of nine patients (44%) in the rate control group undergoing AV valve operation.

**TABLE 3** Baseline echocardiographic characteristics

| | Rhythm control (*n* = 9) | Rate control (*n* = 15) | *P* value |
|---|---|---|---|
| Left atrial size[a] | | | .3 |
| Normal | 3 (37.5) | 1 (10.0) | |
| Mildly dilated | 0 | 3 (30.0) | |
| Moderately dilated | 2 (25.0) | 3 (30.0) | |
| Severely dilated | 3 (37.5) | 3 (30.0) | |
| Right atrial size[a] | | | .4 |
| Normal | 0 | 1 (10.0) | |
| Mildly dilated | 0 | 2 (20.0) | |
| Moderately dilated | 2 (25.0) | 3 (30.0) | |
| Severely dilated | 6 (75.0) | 4 (40.0) | |
| LV ejection fraction (%) | 54.4 ± 12.4 | 60.4 ± 11.1 | .3 |
| LV end-diastolic diameter (cm) | 4.6 ± 0.4 | 5.0 ± 0.4 | .09 |
| RV systolic function | | | .2 |
| Normal | 1 (11.1) | 4 (26.7) | |
| Mildly reduced | 2 (22.2) | 7 (46.7) | |
| Moderately reduced | 3 (33.3) | 1 (6.7) | |
| Severely reduced | 3 (33.3) | 3 (20.0) | |
| RV size | | | .7 |
| Normal | 0 | 1 (8.3) | |
| Mildly enlarged | 1 (14.3) | 2 (16.7) | |
| Moderately enlarged | 4 (57.1) | 3 (25.0) | |
| Severely enlarged | 2 (28.6) | 6 (50.0) | |
| AVVR | | | |
| Affected valve morphology | | | .9 |
| Tricuspid | 7 (77.8) | 12 (80.0) | |
| Mitral | 2 (22.2) | 3 (20.0) | |
| Affected valve position | | | .7 |
| Subpulmonic | 6 (66.7) | 8 (53.3) | |
| Subsystemic | 3 (33.3) | 7 (46.7) | |
| Severity | | | .9 |
| Moderate-to-severe | 5 (55.6) | 8 (53.3) | |
| Severe | 4 (44.4) | 7 (46.7) | |
| Severe aortic regurgitation | 1 (11.1) | 0 | .4 |
| Severe pulmonic regurgitation | 0 | 1 (6.7) | .9 |

*Note:* Values are *n* (%) or mean ± SD.
Abbreviations: LV, left ventricle; RV, right ventricle; AVVR, atrioventricular valve regurgitation.
[a]Atrial size not measured in patients with intra-atrial baffle.

**TABLE 4** Follow-up echocardiographic characteristics

| | Rhythm control (*n* = 9) | Rate control (*n* = 15) | *P* value |
|---|---|---|---|
| Left atrial size[a] | | | .9 |
| Normal | 3 (37.5) | 3 (30.0) | |
| Mildly dilated | 1 (12.5) | 1 (10.0) | |
| Moderately dilated | 1 (12.5) | 2 (20.0) | |
| Severely dilated | 3 (37.5) | 4 (40.0) | |
| Right atrial size[a] | | | .8 |
| Normal | 0 | 1 (10.0) | |
| Mildly dilated | 1 (12.5) | 1 (10.0) | |
| Moderately dilated | 2 (25.0) | 1 (10.0) | |
| Severely dilated | 5 (62.5) | 7 (70.0) | |
| LV ejection fraction (%) | 60.0 ± 11.5 | 58.9 ± 9.0 | .8 |
| LV end-diastolic diameter (cm) | 4.6 ± 0.5 | 4.9 ± 0.7 | .4 |
| RV systolic function | | | .8 |
| Normal | 3 (33.3) | 4 (26.7) | |
| Mildly reduced | 2 (22.2) | 5 (33.3) | |
| Moderately reduced | 3 (33.3) | 3 (20.0) | |
| Severely reduced | 1 (11.1) | 3 (20.0) | |
| RV size | | | .7 |
| Normal | 0 | 1 (12.5) | |
| Mildly enlarged | 2 (28.6) | 1 (12.5) | |
| Moderately enlarged | 3 (42.9) | 4 (25.0) | |
| Severely enlarged | 2 (28.6) | 6 (50.0) | |
| AVVR | | | |
| Severity | | | .9 |
| Mild-to-moderate | 2 (22.2) | 0 | |
| Moderate | 1 (11.1) | 3 (20.0) | |
| Moderate-to-severe | 2 (22.2) | 7 (46.7) | |
| Severe | 4 (44.4) | 5 (33.3) | |

*Note:* Values are *n* (%) or mean ± SD.
Abbreviations: LV, left ventricle; RV, right ventricle; AVVR, atrioventricular valve regurgitation.
[a]Atrial size not measured in patients with intra-atrial baffle.

## 4 | DISCUSSION

In our cohort of 24 ACHD patients with atrial arrhythmias and at least moderate-severe AVVR, attempted rhythm control was associated with greater freedom from AV valve operation and transplant. Patients undergoing attempted rhythm control experienced a reduction in arrhythmia burden and increase in biventricular systolic function. This improvement in ventricular systolic function seemed to postpone the need for surgery, despite unchanged severity of AVVR.

Atrial arrhythmias are common among ACHD patients and often challenging to manage. Previous studies have reported modest success with antiarrhythmic drugs and slightly better results with catheter ablation.[8,10,21] These findings likely reflect the challenges of arrhythmia interventions in a population with a high burden of atrial scar, chamber enlargement, and elevated pressure secondary to AVVR. Nonetheless, 66.7% of patients in the rhythm control group achieved a reduction in arrhythmia burden, as defined by no recurrences or decrease from persistent at baseline to paroxysmal on treatment. It is important to note that reduction in arrhythmia burden appears sufficient to improve ventricular systolic function. This is analogous to premature ventricular contraction (PVC) induced cardiomyopathy among adults without congenital heart disease, where

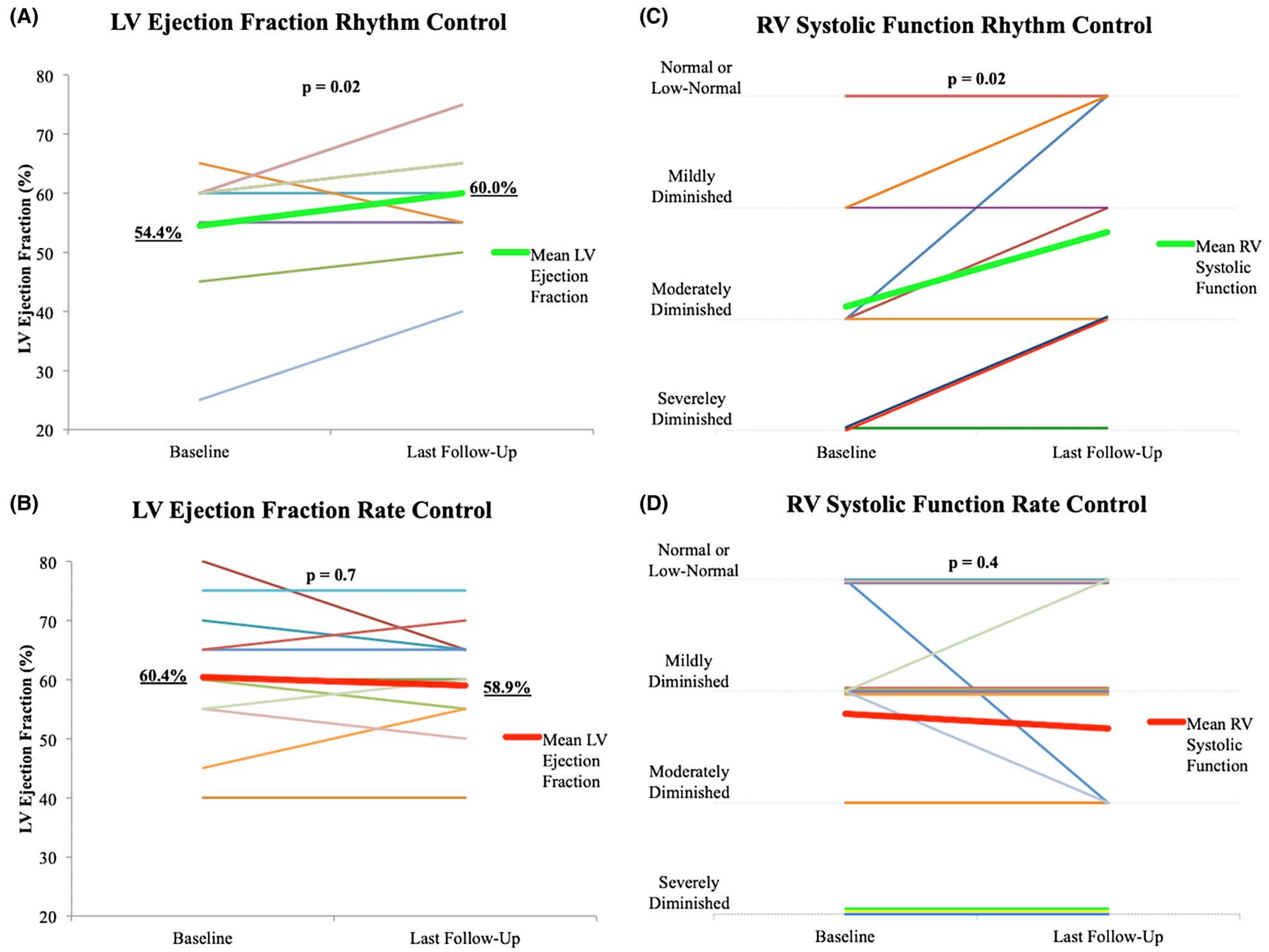


**FIGURE 2** Left ventricular ejection fraction and right ventricular systolic function at baseline and last follow-up among the rhythm and rate control groups. Panel A: Left ventricular (LV) ejection fraction improved significantly from baseline to last follow-up among the rhythm control group ($P = .02$). Each patient is represented as an individual line. The thick line represents mean values. Panel B: LV ejection fraction did not improve among the rate control group ($P = .7$). Panel C: Right ventricular (RV) systolic function improved significantly from baseline to last follow-up among the rhythm control group ($P = .02$). Panel D: RV systolic function did not improve among the rate control group ($P = .4$)

an 80% reduction in PVC burden is generally sufficient to reverse the cardiomyopathy.[22]

Interestingly, the decrease in AV valve operation did not result from decrease in AVVR, but rather from improvement in ventricular systolic function. This finding is in contrast to a previous report of reduction in mitral regurgitation following successful ablation of atrial fibrillation in adults without congenital heart disease.[17] Certainly, rhythm control would not be expected to fix mechanical causes of AVVR, including flail leaflets or impingement by transvenous pacemaker or defibrillator leads.[23] Nevertheless, by reducing atrial arrhythmias and improving ventricular systolic function, the overall improvement in cardiac performance and symptoms seems to have been sufficient to delay the need for surgery.

ACHD providers are frequently tasked with caring for patients with concurrent atrial arrhythmias and AVVR. AV valve surgery is among the most common reoperations in ACHD, and is associated with significant morbidity and mortality.[24,25] These reoperations also contribute substantially to health care resource utilization in the ACHD population.[3,26,27] Our series suggests that a trial of rhythm control may be a reasonable alternative to immediate surgical referral in this patient population.

There are several limitations to our study. Patients were not randomized to treatment strategy, and thus we report an association but cannot establish causality. Patients treated with rhythm control were more likely to have IART than patients treated with rate control. Additionally, there was a nonsignificant trend toward higher rates of persistent atrial arrhythmias among those treated with rhythm control. A higher burden of more easily treated arrhythmia may have influenced the treatment toward rhythm control. Conversely, there was a nonsignificant trend toward higher rates of medical comorbidities among the rate control group, which may have biased outcomes unfavorably. Given our limited sample size, we may have failed to detect true differences in measured baseline characteristics. Further, the possibility of residual confounding by unmeasured baseline characteristics remains.

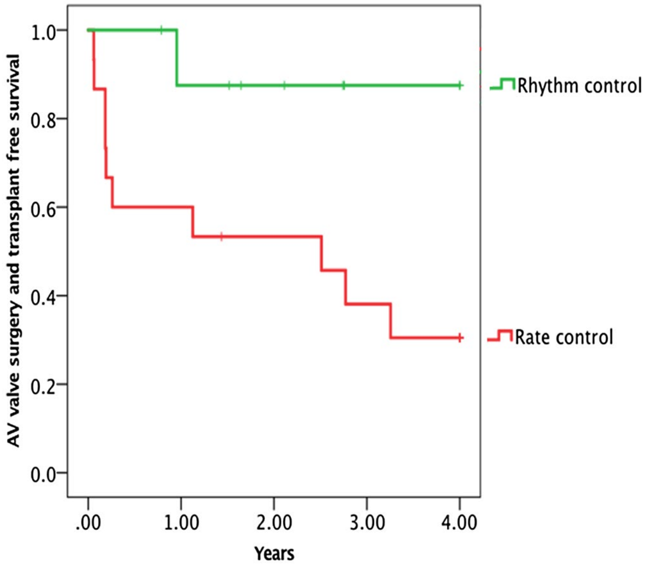


**FIGURE 3** Survival free of AV valve surgery and heart transplant. Survival free of AV valve surgery and heart transplant was greater in the rhythm control group than the rate control group (Log-rank *P* = .04)

Therefore, multicenter studies of larger size are warranted to confirm our findings. While our subjects had a broad variety of congenital lesions, none had single ventricle physiology, which prior studies have identified as particularly resistant to rhythm control.[12] Lastly, as a tertiary referral center for both ACHD and cardiac electrophysiology, our outcomes may not be applicable to other health care settings.

## 5 | CONCLUSIONS

Attempted rhythm control of atrial arrhythmias is associated with improved biventricular systolic function among adults with congenital heart disease and moderate-severe AVVR. This improvement in ventricular function and symptoms may allow surgery to be deferred. Our study supports an initial attempt at rhythm control among such patients, prior to referral for AV valve surgery. Additional studies are needed to further elucidate the relationship between atrial arrhythmias and valvular function among patients with ACHD.

### CONFLICT OF INTEREST

None.

### AUTHOR CONTRIBUTIONS

*Contributed to the study concept and design, data collection and analysis, and drafting of the article*: B Zielonka and DS Frankel
*Contributed to concept and design, and data collection and analysis*: YY Kim
*Contributed to data interpretation and analysis*: GE Supple, SL Partington, ES Ruckdeschel, and FE Marchlinski
All authors provided critical revisions of the submitted article for intellectual content. All authors provided final approval of the article to be published.

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## SUPPORTING INFORMATION

Additional supporting information may be found online in the Supporting Information section at the end of the article.

**How to cite this article:** Zielonka B, Kim YY, Supple GE, et al. Improvement in ventricular function with rhythm control of atrial arrhythmias may delay the need for atrioventricular valve surgery in adults with congenital heart disease. *Congenital Heart Disease*. 2019;14:931–938. https://doi.org/10.1111/chd.12833