Received: 21 June 2018 | Revised: 7 October 2018 | Accepted: 29 November 2018

DOI: 10.1111/chd.12736

ORIGINAL ARTICLE

WILEY Congenital Heart Disease

Extended cardiac ambulatory rhythm monitoring in adults with congenital heart disease: Arrhythmia detection and impact of extended monitoring

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Funding information
This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Abstract

Background: Arrhythmias are a leading cause of death in adults with congenital heart disease (ACHD). While 24-48-hour monitors are often used to assess arrhythmia burden, extended continuous ambulatory rhythm monitors (ECAM) can record 2 weeks of data. The utility of this device and the arrhythmia burden identified beyond 48-hour monitoring have not been evaluated in the ACHD population. Additionally, the impact of ECAM has not been studied to determine management recommendations.

Objective: To address the preliminary question, we hypothesized that clinically significant arrhythmias would be detected on ECAM beyond 48 hours and this would lead to clinical management changes.

Methods: A single center retrospective cohort study of ACHD patients undergoing ECAM from June 2013 to May 2016 was performed. The number and type of arrhythmias detected within and beyond the first 48 hours of monitoring were compared using Kaplan-Meier curves and Cox proportional hazard models.

Results: Three hundred fourteen patients had monitors performed [median age 31 (IQR 25-41) years, 61% female). Significant arrhythmias were identified in 156 patients (50%), of which 46% were noted within 48 hours. A management change based on an arrhythmia was made in 49 patients (16%).

Conclusions: ECAM detects more clinically significant arrhythmias than standard 48-hour monitoring in ACHD patients. Management changes, including medication changes, further testing or imaging, and procedures, were made based on results of ECAM. Recommendations and guidelines have been made based on arrhythmias on 48-hour monitoring; the predictive ability and clinical consequence of arrhythmias found on ECAM are not yet known.

KEYWORDS
adult congenital heart disease, arrhythmia, cardiac rhythm monitoring, screening

1 | INTRODUCTION

Arrhythmias are the leading cause of hospital admission and one of the leading causes of death in adults with congenital heart disease (ACHD).[1-3] Arrhythmia burden in this population has been shown to increase significantly with age, and arrhythmia detection is essential to management and risk assessment.[1] The 2008 ACC/AHA Guidelines for the Management of ACHD recommend selective screening for arrhythmias based on anatomic diagnosis and history.[3] The main tool that has historically been utilized for arrhythmia detection and screening is the noninvasive ambulatory ECG monitor, or Holter monitor, which can record 24-48 hours of electrocardiographic activity.[4] Adhesive patch extended cardiac ambulatory monitoring (ECAM) devices (Zio monitor, iRhythm, San Francisco, California) are now available and can record continuous single lead ECG data for up to 14 days.[4] Several recent studies in adults have demonstrated that ECAM can identify more arrhythmias than standard 48 hours of Holter monitoring.[4,5] The utility of this device and the arrhythmia burden identified beyond 24-48 hours of monitoring have not been evaluated in the ACHD population. Additionally, the impact of the use of ECAM and the utility of the data have not been studied to determine management recommendations based on these results.

The purpose of this study was to investigate the clinical utility of ECAM and to determine if clinically significant arrhythmias would be identified by extended monitoring in ACHD. The primary aim was to determine if arrhythmia monitoring for >48 hours would identify more clinically significant arrhythmias than typical 24-48 hours of monitoring in ACHD patients. We also sought to determine if identification of arrhythmias by ECAM led to changes in patient management and to identify factors associated with arrhythmia development in this population. There are currently no management guidelines using ECAM for arrhythmia diagnosis in ACHD patients. The information in this study will hopefully be a stepping stone to determine how ECAM will help to shape future management guidelines. To address the preliminary question, we hypothesized that clinically significant arrhythmias would be detected on ECAM beyond 48 hours of monitoring and that use of ECAM would lead to changes in clinical management in ACHD patients.

2 | METHODS

This was a single center, retrospective cohort study of ACHD patients followed at the Adult Congenital Heart program at Stanford between June 2013 and May 2016. All patients over 18 years of age with congenital heart disease (CHD) who underwent ECAM with a Zio monitor (iRhythm, San Francisco, California) were included. Patients were excluded if they only had an alternative type of monitor performed. The Zio monitor is an ECG monitor that adheres to the chest and records and stores up to 14 days of continuous ECG. Patients can press a button on the device and fill out a log to document symptomatic events during their wear. After a patient completes the recording, the monitor is mailed to iRhythm Technologies, Inc, where the recording is analyzed using a combination of proprietary algorithms and review by Certified Cardiac Technicians. The findings are then reported to the ordering physician in a report that includes information on several standard arrhythmias, including atrial fibrillation and flutter, ventricular tachycardia, supraventricular tachycardia, atrioventricular pauses, heart block, and atrial and ventricular ectopic beats. Further details on the Zio monitor and its analytic algorithms have been described previously.[5,9] The Zio monitor was the predominant rhythm monitor used by all ACHD providers during this time period and very few patients received a standard Holter or event monitor. The study was approved by the institutional review board at Stanford University.

The following information was retrospectively collected on each patient by chart review: age, gender, body mass index, race, baseline oxygen saturation, baseline EKG including rhythm, rate, corrected QT interval (QTc), and QRS duration, type of CHD, surgical history (including type and date), prior history of arrhythmias, presence of an implanted cardioverter/defibrillator or pacemaker, medications, prior electrophysiology study and ablation procedure, patient symptom history, and echocardiographic findings (including left and right ventricular function, atrial dilation, and estimated right ventricular systolic pressure). Data collected from the ECAM included: the reason for obtaining the ECAM, when arrhythmias occurred (<24 hours, 24-48 hours, >48 hours of monitor start date), type and burden of arrhythmias noted, whether arrhythmias were sustained or nonsustained, and duration of time the monitor was worn. Timing of arrhythmias, in regards to <24 hours, 24-48 hours, and >48 hours, refers to time after the monitor was activated on placement. Follow-up data included clinical management changes performed as a result of the ECAM, including cardiac surgery, electrophysiology study, cardiac catheterization, imaging (echocardiography, cardiac magnetic resonance imaging, computed tomography), medication changes, pacemaker or ICD implantation or re-programming, hospitalization, or cardioversion.

The raw data from each Zio monitor was obtained from iRhythm. The accuracy and validity of the data were determined by over-reading the rhythm strips from a random sample of 10% of monitors by study principal investigator to ensure agreement between iRhythm data, arrhythmia over-readers (ACHD and ACHD EP teams), and the research team. No changes were made to final reads of any of the rhythm monitors in this 10%, and so no further over-reading was done.

Clinically significant arrhythmias were defined as any of the following: atrial fibrillation or flutter (AF), ventricular tachycardia (VT), nonsustained VT (NSVT), supraventricular tachycardia (SVT), sinus pause >3 seconds, second-degree type II atrioventricular block (AVB), and third-degree AVB. Atrial fibrillation was diagnosed when occurring for >30 seconds. Atrial fibrillation is defined with duration >30 seconds in the HRS/EHRA/ECAS expert consensus statement.[6] Shorter episodes of atrial fibrillation may be included in "SVT." SVT was defined as occurrence of 4 or more consecutive beats of ectopic atrial tachycardia or reentrant SVT. NSVT was defined as VT for 4 or more beats and <30 seconds in duration. Sustained VT was defined as VT > 30 seconds in duration.

Indications for monitoring were classified as (1) known history of an arrhythmia prior to ECAM (such as NSVT or SVT) which was previously diagnosed by inpatient or outpatient monitoring, (2) patient symptoms such as palpitations, chest pain, dizziness, or shortness of breath, (3) abnormal testing prior to monitor, and (4) screening. The indication for a monitor was defined as "screening" if the monitor was ordered in an asymptomatic patient with no prior arrhythmia history and no other abnormal testing concerning for an arrhythmia. Symptom and screening categories were defined prior to data collection and analysis.

To determine if an arrhythmia detected on ECAM affected clinical management, the following factors were determined a priori and considered significant changes to patient care if they were noted in the medical record to be made based on Zio patch data: start, termination, or dose adjustment of an antiarrhythmic or other cardiac medication; invasive electrophysiology testing and/or ablation; insertion, removal, or reprogramming of a permanent pacemaker or ICD; cardiac surgery; cardiac catheterization; cardiac imaging (echo, CT, or MRI); hospitalization; or cardioversion.

The primary endpoint was time to first arrhythmia detected on continuous monitoring, whether prior to or after 48 hours of monitoring. The secondary end point was whether management changes were made based on arrhythmias detected on ECAM. When patients had more than one monitor performed, only the first monitor was analyzed for statistical clarity. Descriptive statistics were expressed as numbers and percentages for categorical variables, using chi-square tests. For continuous variables, statistics were described as mean ± standard deviation for continuous variables. If the assumption of normality was violated, statistics were described as median with interquartile range (IQR). Age was analyzed using the Mann-Whitney *U* test. We used Cox proportional hazard univariate regressions to assess the association between first arrhythmia and different CHD diagnoses, baseline demographics and characteristics as listed in the data collection section, and the indication for ordering ECAM (ie, screening, symptoms, other abnormal testing, or history of arrhythmia). Those significantly associated were then further assessed in the multivariate Cox proportional hazard model. The assumption of proportional hazard for each variable was checked using shoenfeld residuals.

Kaplan-Meier curve data were evaluated to determine if clinically significant arrhythmias were identified on ECAM that would not have been identified by the standard 24-48 hours of monitoring period, and to show freedom from arrhythmia based on age, anatomic diagnosis, and indication for monitoring. Arrhythmia timing (either within 48 hours or on full monitor) and monitoring indication were analyzed and assessed with whether or not care changes were made. Two-sided *P* values <.05 were considered statistically significant. Statistical analyses were performed using SAS software, version 9.4 (SAS Institute, Cary, North Carolina).

3 | RESULTS

A total of 382 ECAM were performed in 314 patients (median age 31 [25-41] years, 61% female). Seven charts were not able to be accessed, so baseline data was analyzed for the remaining 307 patients. ECAM data was available for all 314 patients. The 10% of ECAM that were over-read to ensure accuracy of the data showed no discrepencies between the initial read and the overread. Baseline patient characteristics and the indication of symptoms for ECAM are reported in Table 1. The most common indication for ECAM was patient reported symptoms in 39% of patients. The indication of symptoms was examined separately and compared with other indications (arrhythmia history, screening, or abnormal testing) as this is a very common use of the monitors and represents a clinical scenario in which the etiology of a patient symptom must be identified or explained. Patients wore the Zio monitor for an average of 9.5 ± 4.1 days.

The most common diagnoses included: tetralogy of Fallot, atrial septal defect (ASD) and/or partial anomalous pulmonary venous return (PAPVR), single ventricle, D-transposition of the great arteries (D-TGA), and left heart obstructive disease. Pulmonary hypertension associated with CHD was noted in 25 patients (8%).

A significant arrhythmia history was noted in 105 patients (33%), with 32 (10%) having atrial fibrillation, 44 (14%) atrial flutter, 34 (11%) nonsustained ventricular tachycardia, and 33 (11%) supraventricular tachycardia. The most common cardiac medications included beta blockers, ACE inhibitors or angiotensin blockers, diuretics, digoxin, amiodarone, and other antiarrhythmics (see Table 1 for number of patients and percentages).

The indications for ECAM included history of arrhythmia (62, 20%), symptoms (121, 39%), and screening for arrhythmias due to a history of CHD (85, 28%). Thirty patients (10%) had both symptoms and history of arrhythmia, and 12 (4%) had abnormal testing.

Overall, 156 patients (50% of ECAM) showed a significant arrhythmia, but only 72 of those (46%) were during the first 48 hours. SVT was the most common arrhythmia, followed by NSVT. NSVT was detected by ECAM in 35 patients (11% of total monitors); only 14 of those (40%) were within 48 hours. SVT was detected by ECAM in 110 patients (35% of total monitors); only 51 of those (46%) were within 48 hours. Four patients had a sinus pause >3 seconds. Three patients had their first pause noted after 48 hours (these patients carried diagnoses of moderate to severe PS, TOF, and Ebstein's anomaly), and 1 patient (who carried a diagnosis of single ventricle) had the first pause noted between 24 and 48 hours.

For total arrhythmias, arrhythmia incidence continued to increase as time went on: 15% at 1 day, 23% at 2 days, 39% at 5 days, 47% at 7 days, 52% at 10 days, and 62% at 14 days. For symptomatic patients, incidence of arrhythmia also continued to increase throughout the 14 days: 12% at 1 day, 18% at 2 days, 36% at 5 days, 43% at 7 days, 50% at 10 days, and 61% at 14 days. Patients who had an arrhythmia on Zio monitoring were significantly older than those who did not have an arrhythmia (median age 33 vs 28 years, $P < .0001$, Mann-Whitney *U* test). Figure 1 shows Kaplan-Meier estimates of freedom from arrhythmias (Figure 1A), stratified by type of arrhythmia (Figure 1B), age (Figure 1C), and indication for monitoring (Figure 1D). As shown in Figure 1C, there was a significant increase in arrhythmia burden with

TABLE 1 Patient characteristics and history

	Total		Zio indication: symptoms		Other Zio indication		*P* value
Number of patients (*n*, %)	314	100%	121	39%	192	61%	
Age, years (IQR)	31	(25,41)	30	(23,39)	32	(26,41)	.19
Female (*n*, %)	190	61%	86	71%	103	53%	**<.01**
Cardiac diagnosis							
Tetralogy of Fallot	61	19%	20	17%	41	21%	.29
D-TGA	32	10%	6	5%	26	14%	**.01**
ASD and/or PAPVR	42	13%	21	17%	21	11%	.11
Single ventricle	38	12%	8	7%	30	16%	**.02**
Pulmonary hypertension	25	8%	7	6%	18	9%	.24
Ventricular septal defect	23	7%	13	11%	10	5%	.07
CC-TGA	15	5%	3	2%	12	6%	.13
Ebstein's anomaly	11	4%	1	1%	10	5%	**.04**
Other RVOT disease	25	8%	13	11%	12	6%	.15
Atrioventricular canal	6	2%	3	2%	3	2%	.56
Left heart obstruction	34	11%	19	16%	15	8%	**.03**
Arrhythmia history							
Any arrhythmia	105	33%	14	12%	91	47%	**<.01**
Atrial fibrillation	32	10%	3	2%	29	15%	**<.01**
Atrial flutter	44	14%	3	2%	41	21%	**<.01**
Nonsustained VT	34	11%	5	4%	29	15%	**<.01**
Reentrant SVT	28	9%	5	4%	23	12%	**.02**
EAT	22	7%	5	4%	17	9%	**.10**
Tachy-brady syndrome	13	4%	2	2%	11	6%	.07
AV block	12	4%	2	2%	10	5%	.11
Prior testing & procedures							
Saturation ≥90%	271	86%	110	91%	161	84%	**.01**
History of heart surgery	244	78%	93	77%	151	79%	.78
History of cardioversion	37	12%	4	3%	33	17%	**<.01**
History of pacemaker	18	6%	3	2%	15	8%	**.04**
History of ICD	11	4%	1	1%	10	5%	**.04**
History of past EP study	44	14%	4	3%	40	21%	**<.01**
History of ablation	24	8%	2	2%	22	11%	**<.01**
Last EKG sinus rhythm	270	86%	116	96%	154	80%	**<.01**
Last EKG QRS > 150	62	20%	17	14%	45	23%	**.04**
Nl syst. vent. funct. (echo)	209	67%	98	81%	111	58%	**<.01**
Medications							
Beta-blocker	65	21%	14	12%	51	27%	**<.01**
ACE inhibitor or ARB	76	24%	20	17%	56	29%	**.01**
Digoxin	20	6%	3	2%	17	9%	**.02**
Aspirin	82	26%	30	25%	52	27%	.6
Diuretic	60	19%	16	13%	44	23%	**.03**
Warfarin	39	12%	7	6%	32	17%	**<.01**
Novel oral anticoagulant	12	4%	2	2%	10	5%	.11
Lovenox	2	1%	2	2%	0	0	.08

(Continued)

TABLE 1 (Continued)

	Total		Zio indication: symptoms		Other Zio indication		*P* value
Amiodarone	15	5%	0	0%	15	8%	**<.01**
Other antiarrhythmic	17	5%	2	2%	15	8%	**.02**
Pulmonary HTN therapy	25	8%	7	6%	18	9%	.24

Number and percentage of total patients with each characteristic, with characteristics broken down by whether the Zio monitor was obtained due to a patient's reported symptoms versus any other indication for rhythm monitoring such as arrhythmia history, abnormal testing, or screening.
[a]Abbreviations: ACE, angiotensin converting enzyme; ARB, angiotensin receptor blocker; ASD, atrial septal defect; AV, atrioventricular; CC-TGA, congenitally corrected transposition of the great arteries; D-TGA, D-transposition of the great arteries; EAT, ectopic atrial tachycardia; EKG, electrocardiogram; EP, electrophysiology; HTN, hypertension; ICD, implantable cardioverter-defibrillator; NSVT, nonsustained ventricular tachycardia; PAPVR, partial anomalous pulmonary venous return; RVOT, right ventricular outflow tract; SVT, supraventricular tachycardia.
The bolding of the column titles was for emphasis. The bolding of the *p* values was meant to emphasize the statistically significant values, but these can be un-bolded.

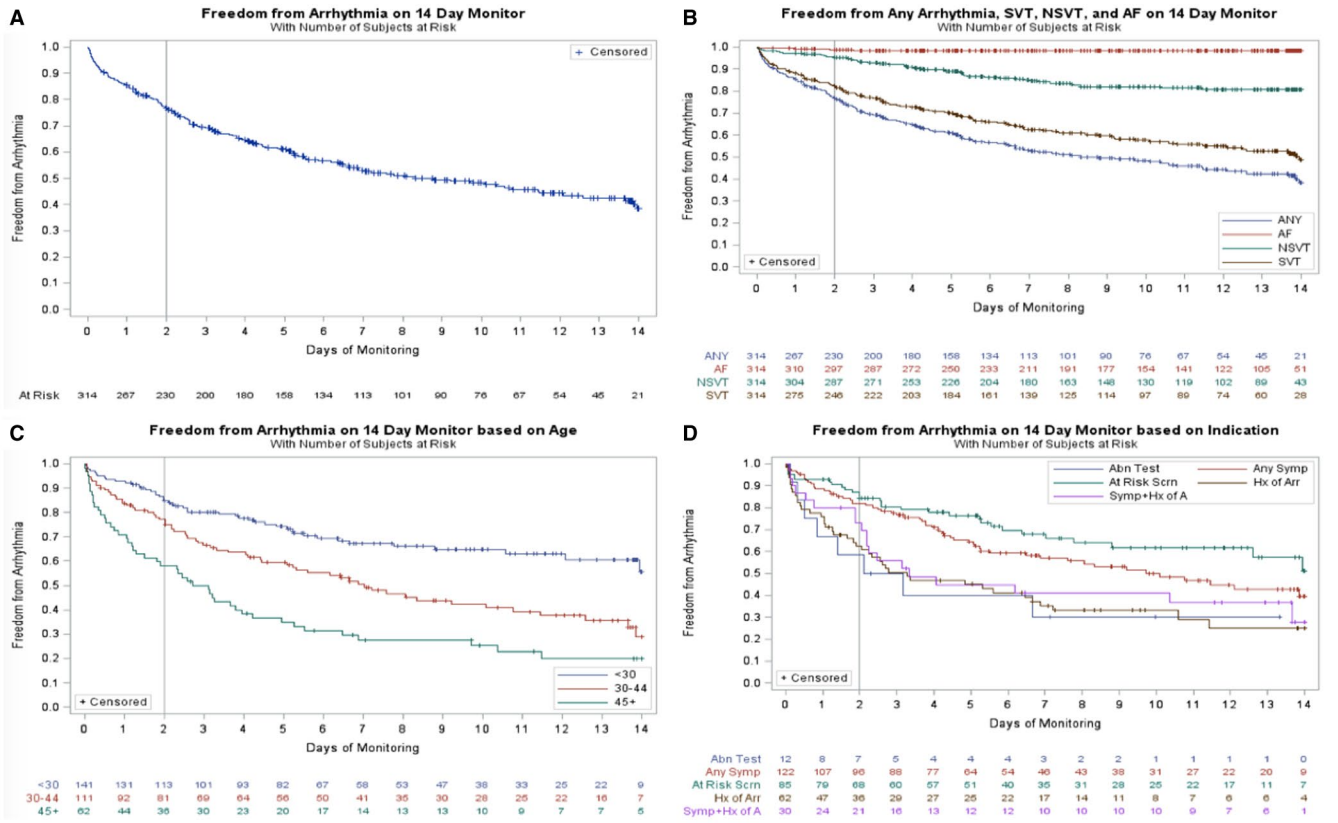

FIGURE 1 Kaplan-Meier Curves of freedom from arrhythmia on all patients (A), based on type of arrhythmia (B), based on age (C), and based on indication for the rhythm monitor (D). A significant arrhythmia burden was noted after the 48-hour mark (the standard monitoring time for the traditional Holter monitor) (A). SVT was the most common arrhythmia identified, followed by NSVT (B). Older patients were more likely to have an arrhythmia (C). Patients with a history of arrhythmia were the most likely to have an arrhythmia noted on monitor, whereas those monitors ordered for screening purposes were the least likely to show an arrhythmia (D)

increasing age ($P < .0001$). As depicted in Figure 1D, patients were most likely to have an arrhythmia noted if the ECAM was ordered due to a history of an arrhythmia; however, an arrhythmia was still detected in 34% of patients who had ECAM for screening purposes. As shown in Figure 2, by completion of the monitoring period, patients with left heart obstructive diseases were most likely to have an arrhythmia ($P = .002$) and patients with tetralogy of Fallot were least likely.

Thirty-seven monitors showed more than one arrhythmia (24% of those with any arrhythmia, 12% of total monitors). Twenty-nine had VT and SVT (19% of monitors with any arrhythmia, 9% of total monitors), 3 had SVT and a sinus pause >3 seconds, 2 had SVT and AV block, and 1 patient each had atrial fibrillation and VT, VT and sinus pause, and VT and AV block. No monitors showed more than two types of arrhythmias. Monitors with two arrhythmias are included in each survival curve based on arrhythmias noted (ie, a monitor showing VT and SVT would show the timing for the first episode each of VT or SVT on the VT or SVT survival curves, respectively, and would reflect just the arrhythmia that occurred first on the "total" or overall freedom from arrhythmia curve).

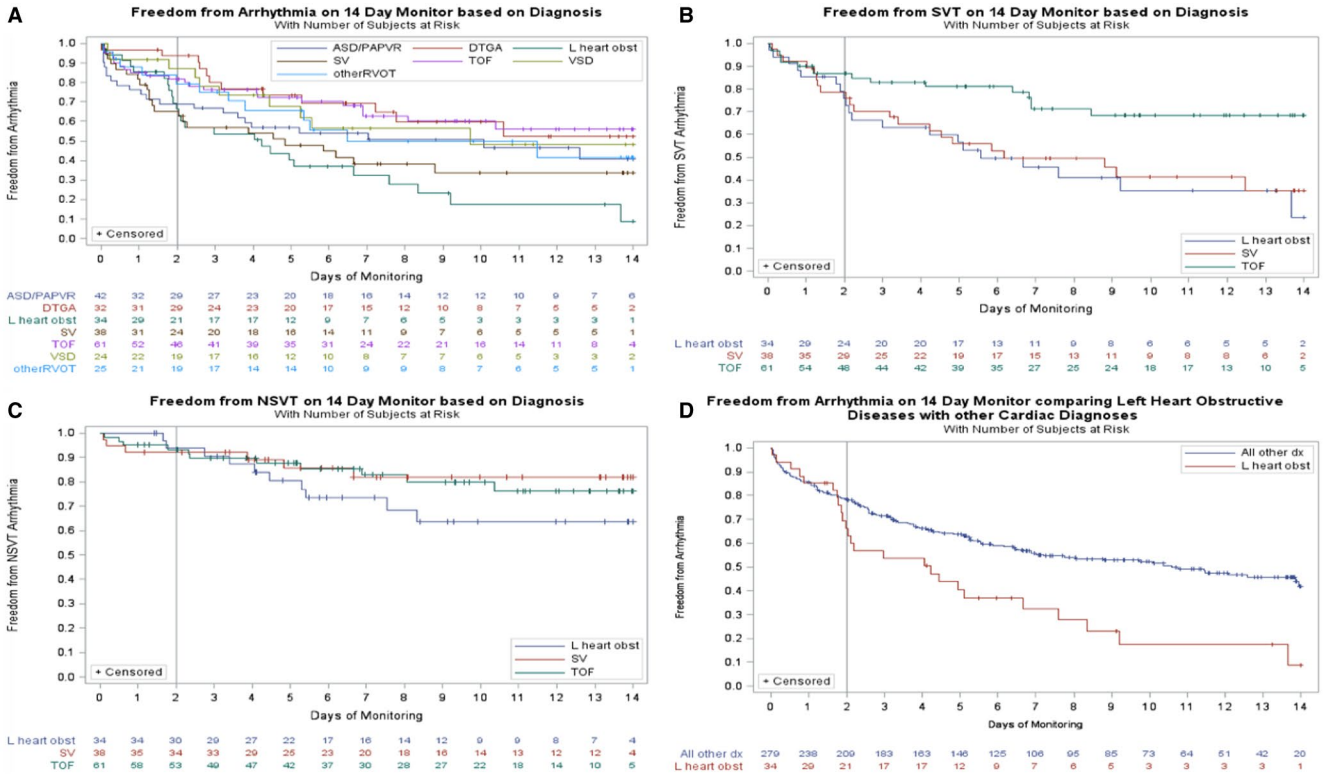

FIGURE 2 Kaplan-Meier Curves based on diagnosis for all arrhythmias (A), SVT (B), and NSVT (C), and all arrhythmias identified in patients with left heart obstructive diseases (D). Patients with left heart obstructive diseases were the most likely to have an arrhythmia. Regardless of diagnosis, a significant arrhythmia burden was noted after monitoring day #2

Patients with left heart obstructive disease were more likely to have SVT and NSVT compared with other diagnoses. On univariate analysis, age, left heart obstructive disease, history of any arrhythmia (atrial fibrillation, atrial flutter, NSVT, SVT, EAT), history of cardioversion, ICD, and treatment with a beta blocker, ACE inhibitor, aspirin, diuretic, novel anticoagulant, and other antiarrhythmic (other than beta-blocker, digoxin, or amiodarone) were significantly associated with arrhythmias on ECAM. In multivariate analysis, age, history of an arrhythmia, left heart obstructive disease, and single ventricle physiology remained significant in association with first arrhythmia observed (Table 2).

As depicted in Figure 3, 49 patients (32%) with an arrhythmia on ECAM had a care change as a result of arrhythmia detection, 28 of whom (9% overall, 57% of care changes) had an arrhythmia noted within 48 hours. An additional 21 patients (7% overall, 43% of care changes) had an arrhythmia after 48 hours that led to a clinical management change. Types of changes made are depicted in Figure 4.

As shown in Figure 5, patients were most likely to have a care change resulting from the rhythm monitor if it was ordered because of a history of an arrhythmia (26%), compared with screening, symptoms, or other abnormal testing. Twenty-three percent of patients who had both symptoms and history of an arrhythmia had a care change. Nineteen percent of monitors ordered for symptoms resulted in a care change, 15% of screening monitors resulted in a care change, and 17% of monitors ordered for other abnormal testing resulted in a care change. Sixteen percent of patients with any arrhythmia had a care change (49 patients). Of the 49 patients with a care change, 57% had care change for arrhythmia noted within 48 hours and 42% had care change for arrhythmia noted after 48 hours. Of the 110 patients with SVT, 37 total care changes were made (34%), and in the 35 patients with NSVT a total of 23 care changes were made (66%).

TABLE 2 Results of multivariate Cox regression analysis assessing factors associated with arrhythmia identification

	Hazard	HR	
Parameter	**Ratio**	**95% CI**	***P* value**
Age	1.03	1.02-1.05	**<.01**
Tetralogy of Fallot	0.84	0.52-1.36	.47
Single ventricle	1.92	1.18-3.11	**.01**
Pulmonary hypertension	1.55	0.93-2.59	.1
Left heart obstructive disease	2.08	1.31-3.30	**<.01**
History of arrhythmia	1.75	1.27-2.43	**<.01**

The bolding of the column titles was for emphasis. The bolding of the *p* values was meant to emphasize the statistically significant values, but these can be un-bolded.

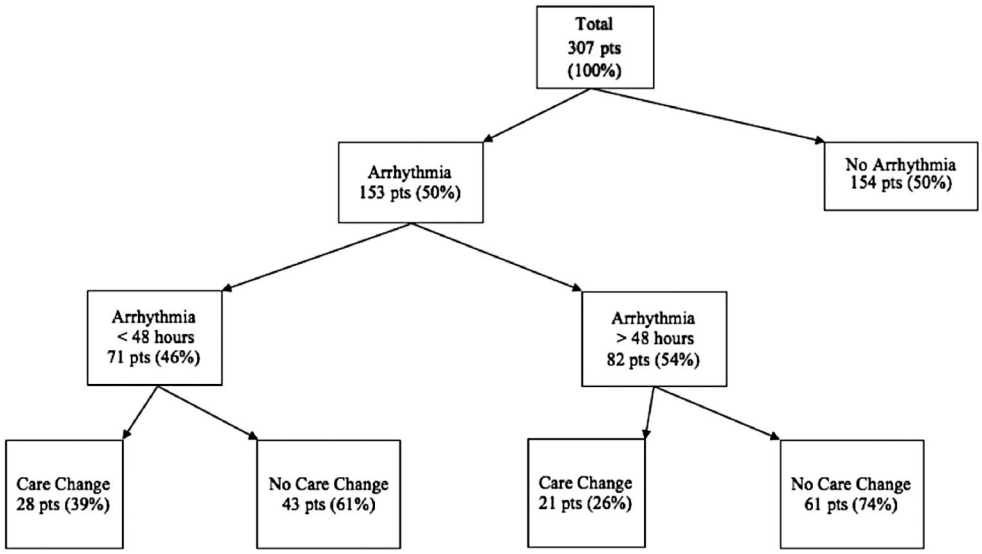

FIGURE 3 Flowchart representation of changes in patient care based on identification of an arrhythmia. Three hundred seven patients were able to be reviewed, of which, 153 had an arrhythmia. Seventy-one patients had the first arrhythmia before 48 hours (46%) and 82 had the first arrhythmia after 48 hours (54%). Of the 49 patients with a care change, 57% had care change for arrhythmia noted within 48 hours, 43% had care change for arrhythmia noted after 48 hours

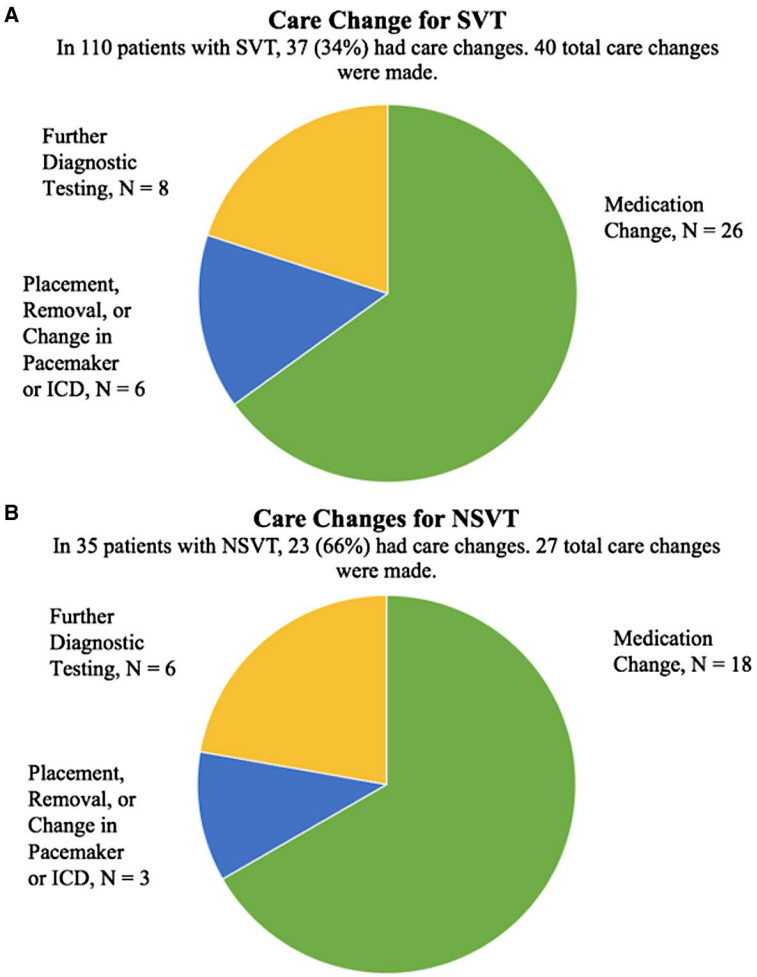

FIGURE 4 Care changes noted for SVT and NSVT. In the 110 patients with SVT, care changes were made in 37 patients (34%). Some patients had more than one care change, totaling 40 care changes (A). In the 35 patients with NSVT, care changes were made in 23 patients (66%). Some patients had more than 1 care change, totaling 27 care changes (B)

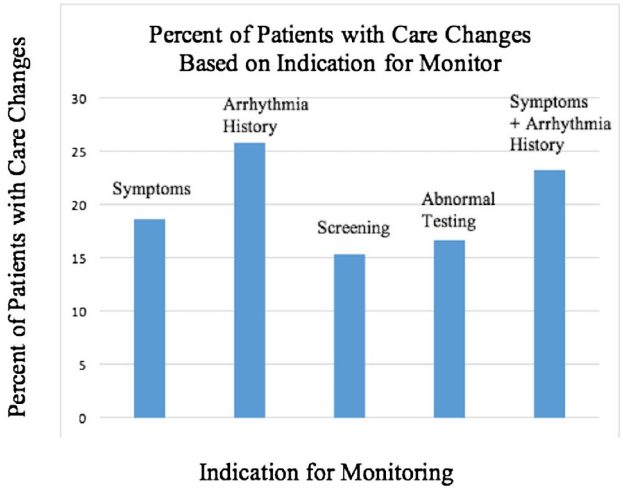

FIGURE 5 Comparison of percent of patients with care changes based on indication for monitor. Care changes were instituted in 19% of patients with monitors ordered for symptoms, 26% of those ordered for history of an arrhythmia, 15% of screening monitors, 17% ordered due to other abnormal testing (EKG), and 23% of those ordered due to both history of an arrhythmia and symptoms

4 | DISCUSSION

With the advent of ECAM devices, patients are routinely being monitored for longer periods of time both to assess symptoms as well as to monitor for occult arrhythmia. While ECAM has been demonstrated to be an effective tool in other populations,[4,7] there has been little data assessing the value and yield in ACHD patients. In this investigation, we found that ECAM for 14 days was more likely to detect arrhythmias than monitoring for 48 hours (as with a typical Holter monitor), and the arrhythmia burden beyond 48 hours of monitoring was substantial. In addition, identification of arrhythmias beyond 48 hours of monitoring directly led to significant care and management changes that may not have occurred with shorter periods of monitoring. As discussed further below, there is little data regarding the significance of arrhythmias noted on extended monitoring, and how this detection should influence patient management.

The burden and impact of arrhythmias in the ACHD population is substantial. Incisional scars, atrial stretch due to hemodynamic impairments, ventricular dysfunction, and other factors make arrhythmias a common occurrence.[1,3] Atrial and ventricular arrhythmias have been shown to occur in up to 43% of patients with tetralogy of Fallot and 100% of patients with Fontan physiology in long term follow-up.[8,9] Single ventricle patients with Fontan palliation have an increased risk of heart transplant, Fontan failure, and death after having their first arrhythmia.[9,10] Arrhythmia detection and identification in this population have important implications for patient management and outcomes.

It seems intuitive that longer monitoring and more detected arrhythmias would result in more changes to patient management. In the symptomatic patient, longer monitoring periods increased the yield of arrhythmia detection (62% of monitors at 14 days vs 23% of monitors at 2 days; impacted medical management in 19% of symptomatic patients). However, the impact of detecting an arrhythmia in an asymptomatic patient is more complicated. We sought to identify whether arrhythmia identification after 48 hours of monitoring directly resulted in management or care changes in asymptomatic patients. In this series overall, for all monitoring indications, ECAM led to a care change in 49 patients, and 21 (43%) of these were for arrhythmias noted after 48 hours. Thus, 43% of care changes were for arrhythmias occurring only after 48 hours.

Changes were clinician dependent, and may have been related to other factors beyond the incidence of an arrhythmia, such as duration and overall arrhythmia burden. Regardless of arrhythmia type, detection of any arrhythmia after 48 hours resulted in care changes in a significant number of patients.

The ability to predict the clinical importance of an arrhythmia identified on ECAM, though, is largely unknown. While NSVT on routine 24-48 hours of Holter monitoring is a marker for sudden death risk in tetralogy of Fallot,[11] it is unknown if that risk extrapolates to the patient with NSVT on day 13 of ECAM. Current recommendations for patients with tetralogy of Fallot and NSVT on a 48-hour Holter monitor may include ICD implantation for an increased risk of sudden cardiac death.[11] It is unknown if escalating therapy based on ECAM is warranted or necessary. It may be the frequency, rather than the initial incidence, of an arrhythmia that is important in making clinical decisions. It will be important to determine the predictive value of this increased detection in future studies. Current guidelines from the Pediatric and Congenital Electrophysiology Society, Heart Rhythm Society, and the European Heart Rhythm Association recommend Holter monitoring for screening and monitoring, and use data generated from Holter monitors as a basis for recommendations.[1,12] Further study will be needed to determine the best management for arrhythmias detected by screening ECAMs, the prognostic utility of arrhythmias on ECAM, and the extent to which they will lead to future life-threatening events. Are 7 episodes of SVT noted on a 14-day extended monitoring equivalent to 1 episode noted on a 48-hours Holter monitor, or is this more significant? Would these two possibilities warrant the same, or differing, interventions? Is 1 episode of NSVT on extended moniting of equal significance to the same episode noted on Holter monitoring, or is one episode on a 14-day monitor less significant? These questions are currently unknown, but very important for the ACHD population.

5 | LIMITATIONS

There were several limitations to this study. First, several diagnostic categories had small patient numbers, thus limiting interpretation of arrhythmia results in these groups. This study also used patients as their own controls, comparing the first 48 hours of ECAM to the full ECAM 14-day recording. While Zio monitors had become the standard monitor ordered in our ACHD clinics during this time, a small number of other monitoring techniques (Holter and event monitors) were also used in select patients. Thus, there may have been some selection bias, with other monitors being used when providers required real-time notification of dangerous arrhythmias. Although the number of such patients was small, their absence from the ECAM cohort

may bias to an underrepresentation of patients in whom significant arrhythmias were detected. In addition, patients were censored for additional episodes of each arrhythmia after the first occurrence of each arrhythmia. The presence of multiple episodes of the same type of arrhythmia was not factored into this analysis. Additionally, for rhythm analysis, if episodes of atrial fibrillation occurred but lasted less than 30 seconds, these would be classified as SVT rather than atrial fibrillation.

There were also limitations to analysis of patient management changes. The impact of de-escalation of care changes (such as stopping an antiarrhythmic) may be challenging, as it was impossible to assess whether this would have occurred with a 48-hours monitor. Finally, patient management decisions were made by individual ACHD providers so variations in practice may be present.

6 | CONCLUSION

ECAM detects more clinically significant arrhythmias than 48 hours of monitoring in ACHD patients. Additionally, ECAM led to potentially important clinical care changes in ACHD patients, including medication changes, further diagnostic testing, and procedures including ICD or PPM placement, removal, or programming change. As arrhythmia detection and management are critical in this unique population, ECAM may be an important tool for screening and clinical care in ACHD patients. Further study is needed, however, to determine what overall risks are present and what management changes are warranted based on ECAM results.

ACKNOWLEDGMENT

The authors would like to thank iRhythm Corporation, including Judy Lenane and Lori Crosson, for assistance with this study.

CONFLICTS OF INTEREST

Data on arrhythmia counts and timing was obtained from iRhythm. Study team members over-read the data on 10% of monitors and agreed with findings. iRhythm had no involvement in hypothesis, study aim generation, or interpretation of results. There was no financial support from iRhythm.

AUTHOR CONTRIBUTIONS

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How to cite this article: Schultz KE, Lui GK, McElhinney DB, et al. Extended cardiac ambulatory rhythm monitoring in adults with congenital heart disease: Arrhythmia detection and impact of extended monitoring. *Congenital Heart Disease*. 2019;14:410–418. https://doi.org/10.1111/chd.12736