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ORIGINAL ARTICLE

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Analysis of adults with congenital heart disease presenting to pediatric emergency departments with arrhythmias

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Abstract

Objective: As survivors of congenital heart disease (CHD) continue to age, healthcare utilization by this population has increased. It is unknown how often these patients utilize the emergency department (ED) at children's hospitals and how arrhythmias play a role in their utilization of care.

Design: Using a retrospective cohort design, the Pediatric Hospital Information System (PHIS) database was investigated and we studied adults (≥18 years) with CHD (ACHD) who presented to pediatric EDs from 2004 to 2014.

Setting: Tertiary care pediatric hospitals.

Results: Of the 6310 encounters to pediatric EDs, 1594 (25%) were for arrhythmias. The median age was 21 years (IQR 19.1–25.1). The most common tachyarrhythmia diagnoses during the study period were atrial flutter (32%), atrial fibrillation (15%), and paroxysmal VT (10%). Bradyarrhythmias represented a minority of total arrhythmias. Presentation with arrhythmias was associated with an increased risk of admission, ICU care, and death ($P < .01$). Arrhythmias are also highly associated with CHD severity with twice as many complex CHD having arrhythmias compared to simple CHD ($P < .01$).

Conclusions: Total admissions of adults with CHD from the ED of children's hospitals have increased over time while the transfers to outside facilities remain consistently low. While the population of ACHD continues to grow, utilization of pediatric EDs for this cohort has increased. Adults with higher CHD complexity are more likely to present with clinical arrhythmias but there is a growing number of ACHD patients with simple complexity presenting with arrhythmias in recent years. ACHD patients that present with arrhythmias are at increased risk of morbidity and mortality.

KEYWORDS
adults, arrhythmias, congenital heart disease

1 | INTRODUCTION

Survivors of congenital heart disease (CHD) continue to increase in the modern era. Due to advances in prenatal diagnosis, palliative surgery and postoperative care, there are more adults surviving with CHD (ACHD) than infants being born with CHD. Over 85–90% of children with congenital heart disease are expected to survive into adulthood.[1] A number of studies of national databases have supported the observation of this trend.[2–4] The number of visits to emergency departments[5] and inpatient hospitalizations[6] are increasing for this subset of the population and these observations represent an evolution in the epidemiology of disease for congenital heart disease.

What was once thought to be a pediatric problem is now becoming burden for adult providers at many levels, primary care and subspecialists alike. A study of the Nationwide Inpatient Sample (NIS) for US demonstrated that the proportion of all ACHD admissions to small/medium-sized hospitals has remained relatively constant despite the growth of self-designated ACHD specialty centers in the United States.[6] This would suggest that there is a major gap in the specialized care required for this patient population. Those without private insurance are at a higher risk of turning to the emergency department for immediate care.[1] The same authors analyzed the National Emergency Department Sample (NEDS) and demonstrated a parallel trend for emergency department visits across the nation for ACHD patients. For both inpatient admissions and emergency room visits for simple and complex congenital heart disease,[5,6] arrhythmias were among the top three reasons ACHD patients present for care.

In the United States, estimates of the true burden of disease are challenging due to a lack of a national health care system and loss of follow up for many patients as they grow out of childhood.[7] Looking at other countries' health care systems, arrhythmias are the most frequent cardiovascular admission diagnosis[2] and increase in prevalence as ACHD patients age.[8–10] A number of database studies have focused on adult hospitals when evaluating the burden of disease in this subset of the population[2,5,6,10] but there have been few studies looking at pediatric hospitals beyond individual centers.

We sought to evaluate the characteristics of ACHD patients presenting to pediatric emergency departments and determine how arrhythmias play a role in their clinical outcomes and test the hypothesis that more ACHD patients with arrhythmias are being transferred out from children's hospitals over time. Using the Pediatric Health Information System (PHIS) database we also characterized the trend in disposition of these patients after presentation to pediatric emergency departments.

2 | METHODS

2.1 | Data source

PHIS is an administrative database[11] that contains data from inpatient, emergency department, ambulatory surgery, and observation encounters from 43 nonprofit tertiary care pediatric hospitals affiliated with the Children's Hospital Association in the United States. The 43 hospitals included in PHIS provide discharge data, including patient demographics, International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) diagnoses, and procedures, length of stay, and discharge status (discharged to home, rehabilitation facility, nursing facility, or inpatient death). Data are de-identified at the time of data submission and are subject to reliability and validity checks before being included in the database.

The database was queried for patients greater than or equal to 18 years of age, carried a diagnosis of congenital heart disease (based on ICD-9 codes) between January 1, 2004 and December 31, 2014 and presented to a pediatric emergency department. The data set was then analyzed using the patients who presented with an arrhythmia at the time of their presentation to the emergency department (ED). The breakdown of arrhythmias was analyzed over the study period and further organized into bradyarrhythmias and tachyarrhythmias. The database also included information on each patient encounter with regard to demographic characteristics, disposition and morbidity and mortality data.

Complexity of congenital heart disease was analyzed using the ICD-9 diagnoses from PHIS and coded based on the AHA/ACC 2008 guidelines for management of adults with congenital heart disease.[12] Patients with arrhythmias were separated based on their complexity and their disposition over time was analyzed using run charts. The PHIS database describes in detail where patients went after their initial presentation to the children's hospital emergency department. The patients discharged from the ER may go home or to a skilled nursing facility or hospice level of care. Likewise, discharge to an inpatient rehabilitation facility can be construed as a transfer. The dispositions were recoded to be clinically meaningful into three main outcomes for disposition: admission, transfer and discharge. A patient encounter was coded as a "discharge" from the ER if they went home, even if under the care of a home health service or self care. If they left the ER and went to another health care facility that was not their individual home, this was considered a transfer.

2.2 | Statistical analyses

All analyses were conducted with STATA version 13.1 (StataCorp. 2013. *Stata Statistical Software: Release 13*. College Station, TX: StataCorp LP). Descriptive statistics were expressed as mean (with standard deviations), median (range and interquartile range [IQR]), and percentages and counts as appropriate. Univariate statistics were analyzed comparing the presence of an arrhythmia to those without arrhythmias in regards to parameters such as ACHD complexity, admission to the hospital, length of stay, ICU admission, need for ECMO and death. Chi squared analyses were carried out in addition to logistic regression to determine odds ratios for determine the risk of arrhythmia with the aforementioned parameters.

3 | RESULTS

The initial data query yielded 6310 patient encounters at the 43 centers. There was an approximate equal representation of men and women (3165 male, 50.2%) and there were 3884 unique patient identifiers suggesting that there was a significant portion of patients (38%) who frequently came to pediatric emergency departments on more than one occasion. The majority of patients was white/Caucasian and of simple complexity based on their CHD lesion. A summary of the demographic characteristics of the initial data query is in Table 1. A comparison of patients with arrhythmias compared to those without is summarized in Table 2.

Of the all the ACHD patients who came to pediatric emergency rooms in the database, 1594 came with a diagnosis of an arrhythmia (25%). Of these patients, the breakdown of arrhythmias is seen in Figure 1. The most common documented tachyarrhythmias were atrial

TABLE 1 Demographic characteristics of ACHD patients presenting to pediatric emergency departments

Total ER ACHD patients	6310
Male ACHD	3165 (50.2%)
Age	21y (25.3–19.1)
White	4555 (72.2%)
Black	1028 (16.3)
Asian	110 (1.7%)
Other	572 (9.1%)
Simple complexity CHD	3765 (59.7%)
Moderate complexity CHD	1198 (18.99%)
Great complexity CHD	1347 (21.4%)
Deaths	68 (1.1%)
ECMO	12 (0.2%)
ICU admission	1059 (16.8%)
Mechanical ventilation	358 (5.7%)
ACHD patients with arrhythmias	1594 (25%)

Values are n, median (interquartile range; range), or % (n).
Abbreviations: ACHD, adult congenital heart disease; CHD, congenital heart disease; ECMO, extracorporeal membrane oxygenation; ER, emergency room; ICU, intensive care unit.

flutter, fibrillation and paroxysmal ventricular tachycardia. The most common bradyarrhythmias were sinus node dysfunction and complete AV block. When looking at CHD severity and the presence of

TABLE 2 Comparisons of those with arrhythmias and those without arrhythmias

	Arrhythmia (n = 1594)	No Arrhythmia (n = 4716)
Admission	1222 (77%)	2138 (45%)
ECMO	6 (0.4%)	6 (0.1%)
ICU admission	506 (32%)	553 (12%)
Mechanical ventilation	163 (10%)	195 (4%)
Deaths	38 (2.4%)	30 (0.6%)
Age	23.2y (20.1–29.6)	20.6y (18.99–23.9)
White	1239 (78%)	3316 (70%)
Black	193 (12%)	835 (18%)
Asian	30 (1.8%)	80 (1.7%)
Simple complexity CHD	766 (48%)	2999 (64%)
Moderate complexity CHD	312 (20%)	886 (19%)
Great complexity CHD	516 (32%)	831 (18%)

Values are n, median (interquartile range; range), or % (n).
Abbreviations: ACHD, adult congenital heart disease; CHD, congenital heart disease; ECMO, extracorporeal membrane oxygenation; ER, emergency room; ICU, intensive care unit.

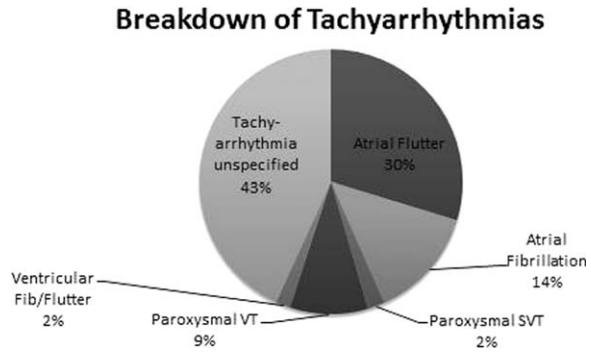

FIGURE 1 Breakdown of tachyarrhythmias of ACHD patients who presented to pediatric emergency rooms 2004–2014

arrhythmia, there is an association between the two variables with an increased risk of arrhythmias with increasing CHD severity. If a patient had CHD of great complexity there was an OR of 2.2 ($P < .01$) of having an arrhythmia compared to CHD of simple complexity (OR 0.5, $P < .01$).

If an ACHD patient came to a pediatric ER, the presence of an arrhythmia was associated with increased morbidity and mortality (Table 2). These patients had a longer length of stay (5.6 vs 3.4 days, $P < .01$), increased risk of ICU admission (32 vs 12%, $P < .01$), mechanical ventilation (10 vs 4%, $P < .01$) and death (2.4 vs 0.6%, $P < .01$). Using logistic regression (Table 3), odds ratios were calculated to determine the odds of these clinical outcomes and the majority of these outcomes were statistically significant ($P < .01$). Having an arrhythmia also increased the odds of being on ECMO, however this was not statistically significant ($P = .06$).

The run chart analyses of ACHD patients' dispositions (Figure 2) demonstrates that over time there has been an increase in admissions and discharges from pediatric emergency departments while the number of transfers has remained relatively unchanged. Thus, the percentage of patients being transferred to other facilities for care has decreased with primary management of the patient occurring at the pediatric hospital of initial presentation. A similar run chart was created focusing on ACHD patients with arrhythmias on presentation to the pediatric emergency departments (Figure 3).

A run chart was created to look at ACHD complexity over time for the study period (Figure 4). The overall complexity of patients has not changed, but there has been a large increase in ACHD patients of simple complexity. A final run chart was created looking at ACHD

TABLE 3 Odds of clinical outcomes associated with arrhythmia

Outcome	OR	*P* Value	95% CI
Admission	3.96	<.01	3.5–4.5
ECMO	2.97	.06	0.96–9.2
ICU admission	3.5	<.01	2.1–3.3
Mechanical ventilation	2.64	<.01	2.1–3.3
Death	3.81	<.01	2.4–6.2

Abbreviations: ECMO, extracorporeal membrane oxygenation, ICU, intensive care unit.

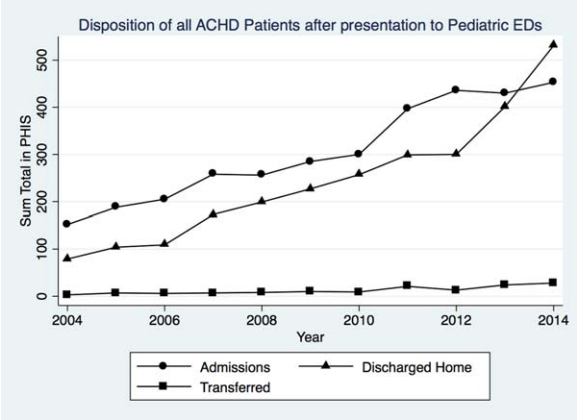

FIGURE 2 Trend in disposition of all ACHD patients after presentation to the pediatric emergency department

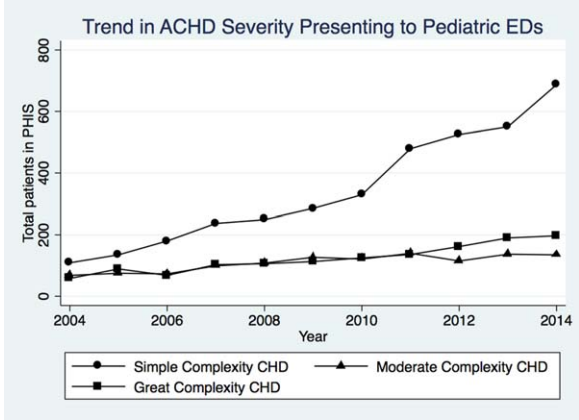

FIGURE 4 Trend in ACHD severity over time

complexity of patients who presented with arrhythmia (Figure 5). Based on these figures, more ACHD patients with arrhythmias are being admitted with the largest proportion coming from those with simple CHD complexity.

4 | DISCUSSION

Our study echoes what has been found in previous studies of national databases looking at the ACHD population.[2,5,6,10] There has been an increase in ACHD ER visits and admissions in recent years in children's hospitals in the United States and arrhythmias are a common problem in this patient population. The proportion of ACHD patients with arrhythmias has remained unchanged over time, although the total amount has increased. Those that present with arrhythmias are at higher risk of morbidity (longer length of stay, ICU admission, higher risk of mechanical ventilation) and mortality. The majority of tachyarrhythmias are atrial flutter/IART or atrial fibrillation, which is what has been reported to be common in this patient population.[8]

Analysis of the trends in ACHD patients with arrhythmias illustrated a number of interesting points. Our hypothesis that more ACHD patients are being admitted and managed at children's hospitals relative to transfers from the ED was not substantiated. More patients are being admitted or discharged over time with the number of transfers remaining relatively unchanged. This would suggest that pediatric hospitals may be becoming more comfortable taking care of ACHD patients due to specialized care teams that are in place. Another possibility is that transition of ACHD patients to adult hospitals has not taken place effectively. This may also be a reflection of the epidemiology of the growing burden of CHD in adults nationally in all adult hospitals in addition to pediatric ones. Agarwal demonstrated this trend in two recent studies looking at national inpatient admissions and ED visits.[5,6] Despite the finding that there is a statistically significant association with CHD severity and arrhythmias in our study, there has been a significant increase in patients with CHD of simple complexity over time (Figure 3). This would suggest that while the risk of arrhythmias increases with CHD complexity, in recent years there is a significant increase in arrhythmias in CHD lesions of simple complexity because many more patients with simple lesions are presenting to care in emergency rooms. While patients with simple complexity may be viewed as less complex, they are at risk of morbidity and mortality from their increasing prevalence of arrhythmias. This may suggest this subgroup requires close outpatient follow up for surveillance of arrhythmias.

Our study has several limitations that can come with large administrative databases. Coding of diagnoses, particularly of arrhythmias and

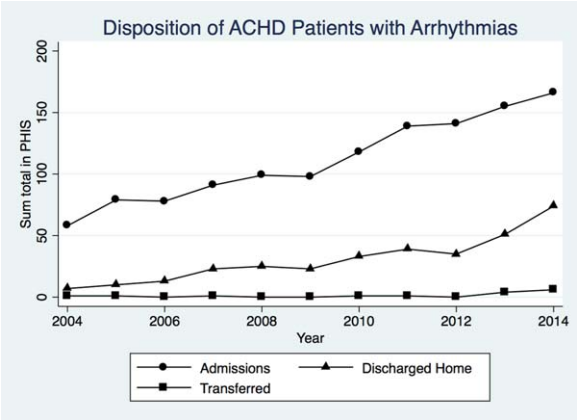

FIGURE 3 Trend in disposition of ACHD patients with arrhythmias from pediatric emergency departments

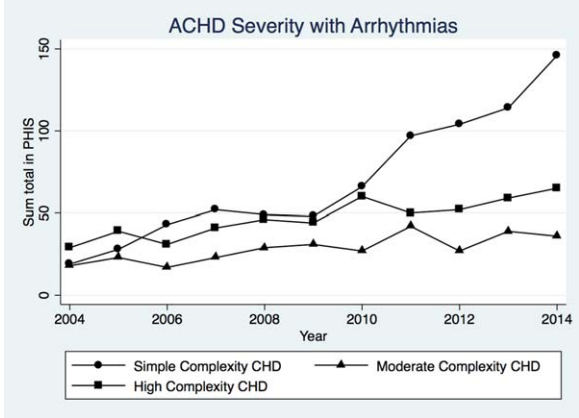

FIGURE 5 Trend in ACHD complexity with arrhythmias over time

CHD lesions, may have inaccuracies.[11] The use of an administrative data set improves the power of our analyses but reliance on the coding of data is dependent on each hospital contributing data to PHIS. The veracity of data is ensured through a joint effort between the Children's Hospital Association and participating hospitals; therefore we rely on the accuracy of the data included from each hospital. Also, we did not specifically look at patients who had more than one visit to the pediatric emergency department and as such the true burden of ACHD patients with or without arrhythmias may be overestimated during the study period.

Our study is unique in that it analyzes the ACHD population that presents acutely to pediatric hospitals in the emergency room setting. As transition of care is one of the major goals for managing patients with congenital heart disease, it will be important to evaluate the trends in both adult and pediatric hospitals as this patient population continues to grow. Our analysis shows an almost fourfold increase in the odds of being admitted to a children's hospital in an ACHD patient if they present with an arrhythmia. Additionally, as might be expected, arrhythmias are associated with both increased morbidity and mortality. Our data shows that there is indeed a discrepancy between admissions and discharges over time for those with arrhythmias. The increase in admissions to and discharges from the primary pediatric centers would suggest that barriers to transition to adult care might still be present despite recommendations for transition starting in childhood.[12] Alternatively, this may be indicative of developing specialized adult congenital programs at pediatric centers, resulting in increased comfort in primary management. Further focused studies in this cohort would be beneficial with particular emphasis on how arrhythmia management may affect outcomes.

AUTHOR CONTRIBUTIONS

Study design: Mohan, Moffett, Lam, Kim
Data analysis/interpretation: Mohan, Moffett, Lam, Kim
Statistics: Mohan, Moffett, Lam, Kim
Drafting the article: Mohan, Moffett, Lam, Kim
Critical revision: Mohan, Moffett, Lam, Kim, Delauz, Miyake, Valdes
Final approval: Mohan, Moffett, Lam, Kim, Delauz, Miyake, Valdes

CONFLICT OF INTEREST

The authors have no disclosures or conflicts of interest.

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