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ORIGINAL ARTICLE

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Long-term follow-up of adult patients with congenital heart disease and an implantable cardioverter defibrillator

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Abstract

Objective: Sudden cardiac death is common in the adult congenital heart disease (ACHD) population. Knowledge and experience about the use of implantable cardioverter defibrillators (ICD) in ACHD patients is very limited. We aimed to characterize a cohort of patients with ACHD and ICDs.

Design: Thirty consecutive ACHD patients submitted to an ICD implantation in a single tertiary center were evaluated. Data on baseline clinical features, heart defect, indication for ICD, type of device, appropriate therapies, ICD-related complication, and mortality during follow-up were collected.

Results: Of the 30 patients, 56.7% received appropriate therapies due to ventricular tachycardia (VT) or ventricular fibrillation (VF). The rate of inappropriate therapies and device-related complications was 33.3%. Secondary prevention and primary prevention patients with class I indications for ICD had more appropriate therapies than complication, but this relationship was reversed for patients with class II indications. Remote monitoring played an important role in diagnosing new atrial arrhythmias before scheduled visits in 46.2% of patients, leading to a change in medication. VT/VF episodes were associated with a composite of death, cardiac transplantation, and hospital admission (OR 13.0; 95% CI: 2.1-81.5).

Conclusion: ICDs are not only useful in preventing SCD, but also have a major role in diagnosing atrial tachyarrhythmias ahead of scheduled visits. Although improvements in ICD technology might reduce complications and inappropriate therapies, adequate selection of candidates for primary prevention still remains difficult because of the lack of clear indications.

KEYWORDS

congenital heart defect, implantable cardioverter defibrillator, supraventricular tachycardia, ventricular fibrillation, ventricular tachycardia

1 | INTRODUCTION

Congenital heart disease (CHD) affects 0.9% of births.[1] With recent advances in clinical management, it is expected that 90% of children with CHD will survive until adulthood,[2] making long-term complication of the disease and its treatment an increasing challenge to the medical community, arrhythmias being especially concerning.[3] Specifically, sudden cardiac death (SCD), mostly caused by sustained ventricular arrhythmias, ie ventricular tachycardia (VT) or ventricular fibrillation (VF), amounts to 20%-25% of all deaths among adult CHD (ACHD) patients.[4,5] However, due to the low prevalence of CHD, patients with such defects represent a minority in the overall population suffering SCD[6] and are therefore underrepresented in clinical trials.

Implantable cardioverter defibrillators (ICDs) have proven to effectively prevent SCD and decrease mortality in patients with acquired cardiomyopathy and are recommended as by international guidelines.[7,8] They have also shown to effectively convert life-threatening arrhythmias in CHD patients,[4] but robust clinical evidence-based guidelines are lacking. In fact, knowledge and experience about the use of ICDs in ACHD is very limited, namely concerning long-term outcomes, as compared with the extensive evidence regarding ICDs follow-up in other groups of patients.

In the present study, we aimed to analyze the circumstances in which ACHD patients received ICDs and to assess outcomes after implantation, including the delivery of appropriate therapies and the rate of complications.

2 | METHODS

2.1 | Selection of patients

A single tertiary referral center's database on ACHD, which amounted to 3684 patients included from 1972 to 2016, was cross-referenced with the same center's ICD database, which included 1049 patients from 1994 to 2016.

2.2 | Cardioverter defibrillator indication and implantation

Indication for ICD were classified as primary or secondary. Secondary prevention included patients who received an ICD after suffering a life-threatening ventricular arrhythmia, that is, sustained symptomatic VT or VF. All other patients were classified as primary prevention and were divided into class I and II indications, according to the most recent guidelines.[9] Cardiac devices were implanted at the discretion of the attending physicians. Parenteral prophylactic antibiotics were administered before the skin incision in all patients. Remote monitoring (RM) was offered to patients whenever available by the device company.

2.3 | Patient evaluation and follow-up

Assessment of ventricular function was performed before ICD implantation. Left ventricular ejection fraction, tricuspid annular plane systolic excursion and tissue Doppler lateral annular tricuspid s' were measured through transthoracic echocardiography. Right ventricular ejection fraction was measured through cardiac magnetic resonance.

Patient follow-up in the device outpatient clinic consisted of a first visit 1 month after implant, followed by regular visits, maximally 6 months apart (1 year apart for patients with RM). Device interrogation was performed in all visits. RM data were transmitted every 3 months and reports were reviewed by trained technicians who would alert the attending physician in case of relevant events. Additionally, an alert-based transmission was performed in response to abnormal events. Patients were then summoned in the following 48 hours for an in-person consultation.

2.4 | Data collection and definitions

Hospital medical records were reviewed to identify baseline characteristics, such as initial diagnosis, previous surgeries, indication for implantable cardioverter defibrillator, type of device and clinical end points. Appropriate therapies were defined as occurring after a device-recorded episode of sustained VT/VF. Device-related adverse events encompass inappropriate therapies and pocket- and lead-related complications. Data regarding arrhythmic events, device complications, and appropriate and inappropriate therapies via ICD was prospectively inputted into a database including all ICD patients. Missing values were then assessed at the time of this study and completed whenever possible. The main measured clinical end point was a composite of all-cause mortality, hospitalization for heart failure or arrhythmia and cardiac transplantation. The components of the primary end point were also individually assessed as secondary end points. Information on hospital admissions and mortality was searched with the use of a nationwide health care platform and systematical revision of patient records. The cause of hospital admission and death was determined according to an International Classification of Disease-9–based system as coded by the discharging hospital.

2.5 | Statistical analysis

Continuous variables are expressed as medians and interquartile ranges. Categorical variables are expressed as frequencies and percentages. Baseline comparisons were performed using the chi-square test for qualitative data and the Student's t test for continuous variables. Univariate regression analysis was used to assess the interaction between the clinical end points and device-related events. A 2-tailed P value <.05 was considered to be statistically significant. All statistical analysis was performed with the software package SPSS, version 23.0 (IBM Corp, Armonk, New York).

3 | RESULTS

A total of 30 ACHD patients with ICD were included. They encompass 0.8% of all ACHD patients and 2.9% of all patients with

ICDs followed in our center. Baseline characteristics at the time of ICD implantation are displayed in Table 1. Median follow-up was 29.7 ± 82.4 months (minimum 6; maximum 234).

3.1 | Congenital heart defects

The most prevalent heart defect was tetralogy of Fallot, followed by *ostium secundum* atrial septal defect and dextro-transposition of the great arteries (Table 2). Eight (26.7%) patients had an additional heart defect and 6 (20.0%) had more than 2 defects. Four (13.3%) patients had pulmonary hypertension and none had Eisenmenger syndrome.

Apart from 1 patient with atrial septal defect and irreversible pulmonary hypertension, all patients were submitted to surgery during childhood and young adulthood (age at first surgery 13.7 ± 27.7 years). Seven (23.3%) patients were submitted to multiple surgeries. Except for the patient with univentricular heart (who had a Fontan surgery) and the 3 patients with *dextro*-transposition of the great arteries (who all had a Rastelli procedure), all other patients had corrective surgery. Median time from surgery to implant was 20.5 ± 17.0 years.

Simple congenital heart defects are not usually associated with the implantation of an ICD, such as atrial septal defect, partial anomalous pulmonary venous drainage, patent ductus arteriosus and pulmonary valve stenosis. The age at first surgery was significantly higher for patients with the aforementioned defects than for patients with more complex heart defects (26.4 ± 54.1 years vs 10.0 ± 27.1 years; $P = .047$). Furthermore, some of these patients had dilated cardiomyopathy (Table 2), which was felt to contribute to the high arrhythmic risk.

TABLE 1 Baseline characteristics

Male gender (*n*, %)	22 (73.3)
Age (years)	39.7 ± 26.1
Hypertension (*n*, %)	5 (16.6)
Diabetes mellitus (*n*, %)	3 (10.0)
Congestive heart failure (*n*, %)	11 (36.7)
Coronary artery disease (*n*, %)	3 (10.0)
Dilated cardiomyopathy (*n*, %)	3 (10.0)
Cerebrovascular disease (*n*, %)	4 (13.3)
Atrial arrhythmia (*n*, %)	7 (23.3)
NYHA class (*n*, %)	
I	11 (36.7)
II	11 (36.7)
III	6 (20.0)
Left ventricle ejection fraction (%)	45.0 ± 26.5
Tricuspid annular plane systolic excursion (mm)	16.0 ± 6.0
Tissue Doppler lateral annular tricuspid s' (cm/s)	9.0 ± 4.0
Right ventricle ejection fraction (%)	32 ± 17
β-blocker (*n*, %)	15 (50.0)
ACE-I/ARB (*n*, %)	14 (46.7)
Antiarrhythmic drug (*n*, %)	19 (63.3)
Anticoagulation (*n*, %)	9 (30.0)

Abbreviations: ACE-I, angiotensin converting enzyme inhibitor; ARB, angiotensin-II receptor blocker; NYHA, New York Heart Association.

3.2 | Implantable cardiac devices

Most patients were fitted with a single-chamber ICD. Four (13.3%) patients had subcutaneous ICDs and 2 (6.7%) patients had cardiac resynchronization therapy (Table 3).

More than half (56.7%) of the ICDs were used for secondary prevention after a VT/VF episode (monomorphic VT: $n = 13$; polymorphic VT: $n = 2$; VF: $n = 2$). Of the patients in the primary prevention group, 7 (53.9%) had a class I indication for ICD and 4 (30.8%) had a class II indication (Table 2). Two (15.4%) patients had no indication for ICD according to current guidelines. These patients had ventricular and atrial septal defects and were judged to have a high arrhythmic risk due to subpulmonary right ventricle systolic dysfunction and nonsustained VT.

3.3 | Implantable cardioverter defibrillator interventions and complications

During follow-up, half of the patients received appropriate ICD interventions due to VT/VF. Median time from implant to first intervention was 21.8 ± 29.1 months. No interaction was found between baseline characteristics or type of heart defect and ICD interventions (*P* value nonsignificant). About 58.8% of patients in the secondary prevention group and 38.5% in the primary prevention group received appropriate interventions. Among the primary prevention group, 57.1% of patients with class I indication and 16.7% with class II indication received appropriate therapies (*P* value nonsignificant for both). Among the class II indications, only 1 patient with tetralogy of Fallot received appropriate therapies. Neither of the 2 patients with no indication received appropriate therapies during a mean follow-up of 6.4 years.

Overall, 10 (33.3%) patients experienced at least 1 adverse event related to the ICD. Six (20.0%) patients suffered inappropriate ICD interventions due to fast conducting supraventricular tachyarrhythmias (atrial fibrillation and flutter) and sinus tachycardia (18.1% in the primary prevention and 25.0% in the secondary prevention group). Four (13.3%) patients were affected by pocket or lead complications requiring reinterventions (15.4% in the primary prevention and 17.7% in the secondary prevention group). Two patients were affected by more than one complication (Table 4). The case of pocket hematoma occurred during the hospital admission for ICD implantation in relation with heparin use, while the other complications occurred after discharge. Among primary prevention patients with non–class I indications, 2 (33.3%) patients suffered device-related adverse events. No interaction was found between the type of heart defect, device or indication, and the occurrence of inappropriate therapies or complications (*P* value nonsignificant).

TABLE 2 Congenital heart defects and distribution of ICD indications

Congenital heart defect	Prevalence (*n* = 30)	Secondary prevention (*n* = 17)	Class I primary prevention (*n* = 7)	Class II primary prevention (*n* = 4)
Tetralogy of Fallot	11 (36.7)	8	1	2
Ostium secundum atrial septal defect[a]	4 (13.3)	1[c]	2[c]	0
Dextro-transposition of the great arteries	3 (10.0)	2	0	1
Ventricular septal defect[b]	3 (10.0)	1	1	0
Aortic coarctation	2 (6.7)	0	1	1
Ebstein disease	2 (6.7)	2	0	0
Partial anomalous pulmonary venous drainage	1 (3.3)	1[c]	0	0
Patent ductus arteriosum	1 (3.3)	0	1	0
Pulmonary valve stenosis	1 (3.3)	1	0	0
Subaortic stenosis	1 (3.3)	1	0	0
Univentricular heart[a]	1 (3.3)	0	1	0

[a]1 patient in this group has pulmonary hypertension.
[b]2 patients in this group have pulmonary hypertension.
[c]1 patient in this group has dilated cardiomyopathy.

TABLE 3 Distribution of implantable cardioverter defibrillator

Single-chamber ICD	19 (63.3)
Double-chamber ICD	5 (16.7)
Subcutaneous ICD	4 (13.3)
CRT-ICD	2 (6.7)

Abbreviations: CRT, cardiac resynchronization therapy; ICD, implantable cardioverter defibrillator.

TABLE 4 Device-related complications

Pocket reintervention	3 (10.0)
Skin adhesions	2 (6.7)
Hematoma	1 (3.3)
Lead replacement due to dysfunction	2 (6.7)
System extraction due to pocket infection	1 (3.3)

3.4 | Arrhythmia detection and treatment

Seventeen (56.7%) patients were on the RM program. Six (46.2%) patients had newly diagnosed atrial arrhythmias through the RM service (atrial flutter: *n* = 2; atrial fibrillation: *n* = 4), resulting in changes in medication (oral anticoagulation and/or antiarrhythmic therapy) for all patients.

Six (20.0%) patients were submitted to percutaneous catheter ablation of macroreentrant tachyarrhythmia (atrial flutter: *n* = 3; VT: *n* = 3). Acute success rate was 100%, but all patients with atrial flutter had recurrences and repeated ablation procedure (median time to redo procedure 118.6 ± 96.0 days).

3.5 | Clinical events

Incidence and annual rates of clinical events are displayed in Table 5. In total, 18 (60.0%) of patients died or were submitted to heart transplant or hospital admission during follow-up, with an annual rate of 7.3%. Secondary prevention patients were more likely to die (85.7% vs 14.3%; *P* = .077). Two cases of sudden death were reported: 1 patient had been submitted to heart transplant with explantation of the ICD 5 years prior; the other did not have RM, but had had previous appropriate ICD interventions due to VT/VF; he and was not submitted to autopsy or postmortem ICD interrogation.

Implantable cardioverter defibrillator interventions were strongly related to a composite of death, transplant, and hospitalization (OR 13.000; 95% CI: 2.074-81.479; *P* = .006), mainly driven by an association to hospitalization (OR 12.375; 95% CI: 1.828-83.767; *P* = .010). No association was found between inappropriate therapies or complications and adverse clinical events.

4 | DISCUSSION

Sudden cardiac death due to sustained ventricular tachyarrhythmias is a well-recognized cause of mortality in ACHD patients.[4,10,11] The main findings of this study were: (1) half of the population of ACHD patients with ICD received appropriate therapies due to VT/VF; (2) the rate of inappropriate therapies and device-related complications was high; (3) RM played an important role in diagnosing atrial arrhythmias; and (4) VT/VF episodes were associated with a composite of death, cardiac transplantation, and hospital admission.

TABLE 5 Incidence and incidence rate of clinical events

Event	Incidence (*n*, %)	Annual rate (%)
Death/transplant/ hospitalization	18 (60.0)	7.3
Death	7 (23.3)	2.8
Transplant	2 (6.7)	0.8
Hospitalization	15 (50.0)	6.1
Hospitalization for heart failure	9 (30.0)	3.6
Hospitalization for arrhythmia	9 (30.0)	3.6

4.1 | Baseline and device characteristics

This cohort is relatively young, with a high prevalence of left and right ventricular dysfunction. Tetralogy of Fallot, transposition of the great arteries and septal defects were the most common congenital heart defects and implantable cardiac device distribution is similar to prior studies.[12] Since this population has low rates of atrioventricular block there is no evidence to support the use of dual-chamber instead of single-chamber ICDs.[13] Little experience is available regarding the use of subcutaneous ICDs in patients with ACHD, but their number is steadily increasing. They are a promising alternative in younger patients with limited venous access to the ventricle or with intracardiac shunts increasing the risk of systemic emboli.[14]

ICD implantation occurred several years after the repair of congenital heart defects. Surgical scars are important substrates for ventricular arrhythmias,[9] but the risk of SCD will only be perceived long after surgery. This emphasizes the importance of the long-term follow-up of these patients. Simple congenital heart defects were corrected later than more complex defects. They have the potential to remain undiagnosed for many years, since they do not initially cause symptoms. With longer times until the correction of the defect, volume overload and dysfunction of the ventricles may ensue, which may not be reversible after surgery.

Dilated cardiomyopathy was a contributing factor to the arrhythmic risk in some patients. Although 10% of patients had coronary artery disease, in none of these patients was it severe enough to cause ischemic cardiomyopathy.[15]

4.2 | Suitability of ICD indication and association with appropriate therapies

Current class I indications for ICD in ACHD are similar to those in acquired cardiomyopathies, for secondary prevention in the case of symptomatic VT/VF and for primary prevention in the case of biventricular physiology, left ventricular ejection fraction <35%, and symptomatic heart failure.[8,9] The decision to implant an ICD after an episode of VT/VF is evident, but prospective evidence for primary prevention in ACHD patients is unavailable. The class II indications are extrapolated from studies in patients with acquired cardiac conditions or derived from retrospective data or expert opinions. They rely on the presence of an indeterminate number of risk factors, have no defined cutoff values for left and right ventricular dysfunction, and, apart from tetralogy of Fallot, are not disease-specific.[9]

This cohort has a very high arrhythmic risk. The rate of appropriate intervention in primary prevention patients is notably higher than in patients with hypertrophic cardiomyopathy,[16] arrhythmogenic right ventricular cardiomyopathy,[17] long-QT syndrome,[18] and ischemic and nonischemic cardiomyopathy.[19-21] It is also higher than in previous studies of ACHD (annual rate 15.6% vs 6.8% for primary prevention and 23.8% vs 8.2% for secondary prevention).[12] This can be explained by 2 probable interfering factors. Firstly, a time bias: with first episodes of VT/VF mostly occurring within 2 years after ICD implantation,[22] studies with longer follow-up will have a lower annual rate of therapies. Secondly, a selection bias toward high-risk patients with a high proportion of class I indications who in all likelihood will have a higher rate of VT/VF episodes. Importantly, the rate of appropriate therapies among primary prevention patients with a class I indication was similar to the rate among secondary prevention patients. This strengthens the conviction that guidelines for the general population can be extrapolated to the population of ACHD with good results.[8]

Only 1 of the patients with a class II indication received appropriate interventions. This may point to potentially unneeded ICD implantations. The exception is the indication regarding tetralogy of Fallot. Because of its high prevalence (7%-10% of all CHDs)[23] and predisposition for ventricular arrhythmias,[24] tetralogy of Fallot is the most studied congenital heart defect in the context of ICD implantation.[12] The disease-specificity of the indication probably results in a higher appropriate intervention rate.[25]

Both nonsustained VT[22] and subpulmonary right ventricle systolic dysfunction[4] have been connected to SCD in ACHD patients. Although these subjects might be perceived as having a high arrhythmic risk, prior reports have already established that this group of patients do not receive appropriate interventions.[12]

4.3 | Burden of inappropriate therapies and device-related complications

High rates of adverse events of ICDs during short- and long-term follow-up have been thoroughly reported.[25-30]

Inappropriate shocks are typically more frequent in the ACHD population than in the general ICD population. The main reasons are: (1) a higher incidence of atrial tachyarrhythmias,[2] (2) a younger age and more active lifestyle leading to sinus tachycardia, and (3) a higher rate of lead dysfunction.[31] The annual rate of inappropriate therapies in our cohort was 8.1% (7.3% in the primary prevention and 10.1% in the secondary prevention group), which is analogous to previous reports.[12] In our series, all inappropriate therapies were triggered by supraventricular tachyarrhythmias, with no oversensing or lead failure.

Unlike previous studies,[32] procedure-related complications during the index hospital admission were rare in our population. However, the overall annual rate of lead- and pocket-related complication requiring repeat intervention was 5.4%. This is consistent with other reports in a similar population,[12] but remarkably higher than ICD recipients with ischemic and nonischemic heart disease.[19] Contributing factors are: (1) the younger age of ACHD patients, with a more active lifestyle; and (2) the need for several generator replacements and additional cardiac surgeries, that can destabilize leads. Concerns about the difficult lead placement in this population with complex anatomy leading to more unstable leads[12] might be unjustified, since the rate of complications is similar to young populations with inherited cardiomyopathies.[18,33,34]

4.4 | Remote monitoring and arrhythmias treatment

RM of cardiac implantable electronic devices provides remote access to device battery, lead parameters and history of arrhythmias.[35] Although there are currently no specific RM trials in the ACHD patients,[36] one could assume that this population would derive at least the same benefit from RM as the general population. RM has already demonstrated to decrease the amount of outpatient and emergency room visits and to improve quality of life.[37,38] A major limitation of conventional outpatient clinic follow-up is the absence of monitoring between hospital visits. Recorded data that could have an impact on morbidity and mortality are either completely missed or remain concealed for extended periods. With high rates of device-related complications,[31] inappropriate therapies,[12] and atrial arrhythmias,[9] these events could be detected earlier and appropriate measures undertaken[37,39] to reduce the number of shocks and spare the device battery.[40] In our cohort, a significant number of patients was diagnosed with atrial fibrillation and flutter before an outpatient visit, which lead to an earlier change in medication. However, it still remains unproven if RM in the ACHD population can decrease death and hospitalizations as for patients with advanced heart failure.[41]

4.5 | Clinical end points

In this population, the annual mortality rate was 2.8%, which is comparable to previous studies in ACHD patients.[12] This rate is higher than in the average population of ACHD patients (2.8% vs 0.8%),[42] likely due to more severe heart disease. However, it is lower than in the ICD population with ischemic and nonischemic cardiomyopathy (2.8% vs 5.8%), owing to younger age at implantation (39.7 vs 60.1 years) and less comorbidities.[19]

Sudden death in congenital heart disease patients can be due to a multitude of causes.[4] In out-of-hospital cardiac arrests, it is often difficult to determine the cause of death. Ventricular tachyarrhythmias are the most frequent cause in this population.[4] However, prolonged asystolic episodes can be involved in situations where the ICD was unable to revert the cardiac arrest. VT storm can also be a cause of sudden death despite multiple appropriate ICD interventions. This can explain why 1 patient in this cohort suffered sudden death despite have a well-functioning ICD.

We found an association between the combined clinical end point death, cardiac transplant and hospitalization and appropriate therapies. Appropriate ICD shocks are a surrogate marker for SCD, but may overestimate the risk for SCD, since some of the ventricular arrhythmias may not be life-threatening.[22] No association was found between inappropriate therapies and complications and clinical adverse events in this cohort, although it has been reported in previous studies.[43-45]

4.6 | Net benefit of implantable cardioverter defibrillator in the ACHD population

ACHD patients are a particular subgroup among patients with ICD, because of the high rate of inappropriate therapies and lead- and generator-related complications.[12] Patients with prior VT/VF episodes are regarded as having a high arrhythmic risk, where ICDs are justified. In primary prevention, there have been concerns that the rate of appropriate therapies would be lower than the rate of inappropriate therapies and complications.[22,46-48] Inappropriate shocks are associated with increased mortality[44,45,49] and major psychological impact in the general ICD population. In ACHD, they negatively affect quality of life, sexual function and social interactions.[50] Moreover, complications are also associated with short- and long-term morbidity and mortality.[43,51]

Patients with class I primary prevention indications have a similar incidence of appropriate therapies compared to acquired heart disease, but lower mortality. The benefit of ICD during very long-term follow-up is expected to be much greater in the ACHD population, due to a more favorable appropriate intervention-to-mortality ratio. Besides, the rate of device-related adverse events was lower than the rate of appropriate therapies. However, in patients with non–class I primary prevention indications, this association was reversed due to a low rate of appropriate therapies. The threshold at which a primary prevention ICD should be implanted is not clear and must take into account the observed complication rate. In hypertrophic cardiomyopathy, an ICD is recommend when the 5-year risk of SCD is ≥6% and may be considered when the risk is between 4% and 6%.[52] Risk stratification is far less well established in patients with ACHD. There is an urgent need for randomized controlled clinical trials to define alternative approaches for risk stratification, to refine candidate selection and to establish clearer primary prevention indications.

4.7 | Improving care of ACHD patients with ICDs

Significant improvements in the clinical management of patients with ACHD have been made in the past years. As implantable devices become more frequent in this population, a wider array of therapies emerges. Appropriate patient selection is key to maximize SCD prevention and to ensure low rates of inappropriate therapies and device-related complications.

Subcutaneous ICDs may be especially valuable to avoid lead failure, but still pose the problems of inappropriate therapies and infection.[14] Furthermore, as the device lacks the features of antitachycardia and chronic antibradycardia pacing, its indications are limited.

Individualized ICD settings and programming is of paramount importance to reduce the number of inappropriate shocks. Programming only 1 therapy zone,[35] increasing detection heart rates and detection duration,[13,53] expanding the use of antitachycardia pacing[54] and using discrimination algorithms based on morphology, stability, and onset[55] have all been linked to fewer shocks and might also reduce mortality.

Tailored antiarrhythmic drug therapy is associated with longer times to first shock and reduced number of shocks.[56] Moreover, it has been suggested for patients with tetralogy of Fallot that prophylactic ablation of potential arrhythmogenic isthmuses could prevent primary prevention ICD implantation.[9] This approach has yet to be tested in a randomized prospective study.

RM appears to have an important role in the timely diagnosis of atrial tachyarrhythmias and should be employed in all ACHD patients, since these events are extremely common[9] and could have an impact on prognosis.

4.8 | Limitations

Although data regarding ICDs were prospectively collected, the remaining information is of a retrospective nature. Lower levels of evidence and a higher risk of selection bias, incomplete outcome data, and reporting bias may apply to this study design. More accurate evidence regarding the outcomes of ICD should be collected through large prospective trials to identify definite risks and benefits of ICD.

Due to the small cohort size of this study, the results may not necessarily be reflective of the whole population of ACHD with ICD. However, this represents the entire ICD population among a much larger population of ACHD patients followed at this center. This might also be the reason why no statistically significant associations were found between baseline characteristics and adverse events.

This cohort was collected through the time span of 22 years. Great advances have been made in recent years in the management of this population. The first patients to be included might not be representative of a contemporary population of ACHD with ICDs in terms of rates of appropriate therapies and device-related adverse events.

5 | CONCLUSION

Patients with ACHD and ICDs represent a very small proportion among the whole ACHD population and also among ICD recipients. ICDs are not only useful in preventing SCD, but have a major role in diagnosing atrial tachyarrhythmias ahead of scheduled visits. This cohort had a remarkably high rate of appropriate ICD interventions. In non–class I primary prevention indications, however, this rate was surpassed by the rate of device-related adverse events. Although improvements in ICD technology might reduce complications and inappropriate therapies, adequate selection of candidates for primary prevention still remains difficult because of the lack of clear indications. Prospective research is urgently required to fill this gap and to prepare for a future where ACHD patients will live a long-lasting life.

CONFLICT OF INTERESTS

No conflicts of interest or funding sources to report.

AUTHOR CONTRIBUTIONS

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