International Journal of Cardiology xxx (xxxx) xxx

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Early detection of ventricular arrhythmias in adults with congenital heart disease using an insertable cardiac monitor (EDVA-CHD study)

Rafi Sakhi [1], Robert M. Kauling [1], Dominic A. Theuns [1], Tamas Szili-Torok [1], Rohit E. Bhagwandien [1], Annemien E. van den Bosch [1], Judith A.A.E. Cuypers [1], Jolien W. Roos-Hesselink [1], Sing-Chien Yap *,[1]

Department of Cardiology, Erasmus MC, University Medical Center Rotterdam, Rotterdam, the Netherlands

ARTICLE INFO

Article history:
Received 15 August 2019
Received in revised form 27 December 2019
Accepted 3 February 2020
Available online xxxx

Keywords:
Sudden cardiac death
Risk stratification
Ventricular arrhythmias
Implantable loop recorder
Congenital heart disease

ABSTRACT

Background: Sudden cardiac death (SCD) due to ventricular arrhythmias (VA) is an important mode of death in adults with congenital heart disease (CHD). Risk stratification is difficult in this heterogeneous population. Insertable cardiac monitors (ICM) may be useful for risk stratification. The purpose of the present study was to evaluate the use of ICM for the detection of VA in adults with CHD.

Methods: In this prospective single-center observational study we included consecutive adults with CHD deemed at risk of VA who received an ICM between March 2013 and February 2019. The decision to implant an ICM was made in a Heart Team consisting of a cardiac electrophysiologist and a cardiologist specialized in CHD.

Results: A total of 30 patients (mean age, 38 ± 15 years; 50% male) received an ICM. During a median follow-up of 16 months, 8 patients (27%) had documented nonsustained VA. Of these 8 patients, 3 (10%) received a prophylactic ICD. Furthermore, ICM-detected arrhythmias were present in 22 patients (73%) leading to a change in clinical management in 16 patients (53%). Besides the patients receiving an ICD, 10 patients (33%) had a change in their antiarrhythmic drugs, 6 patients (20%) underwent an electrophysiology study, and 1 patient (3%) received a pacemaker.

Conclusions: The detection of VA by the ICM contributed to the clinical decision to implant a prophylactic ICD. Furthermore, ICM-detected arrhythmias led to important changes in the clinical management. Therefore, long-term arrhythmia monitoring by an ICM seems valuable for risk stratification in adults with CHD.

1. Introduction

Sudden cardiac death (SCD) is an important mode of death in adults with congenital heart disease (CHD) and is mainly driven by ventricular arrhythmias (VA) [1–4]. Identification of patients with CHD at risk for VA is important to determine which patients may benefit from a prophylactic implantable cardioverter-defibrillator (ICD). Risk stratification is hampered by the low predictive value of clinical risk factors [5]. This is not surprising considering Bayes theorem and the low absolute incidence of SCD in adults with CHD [6]. For patients with tetralogy of Fallot there is some guidance on selecting patients for a prophylactic ICD [7,8]. In other CHD lesions, the decision is challenging and the indication for an ICD is largely based on systemic ventricular dysfunction, syncope and/or documented VA. The decision to implant an ICD is also hampered by potential ICD complications, such as shocks, device or lead malfunction, inappropriate shocks, and psychological burden [9–11].

Considering the clinical relevance of documented VA for risk stratification, we adopted a strategy focusing on early detection of VA using insertable/implantable cardiac monitors (ICMs). ICM-detected VA may provide a tipping point in decision-making in patients who are considered at risk of SCD but who do not qualify for an ICD according to current guidelines. Long-term arrhythmia monitoring using ICMs already has an established role in patients with recurrent syncope [12]. In the most recent ESC Syncope guidelines there is an expanding role for ICMs for risk stratification in patients with primary cardiomyopathy or inheritable arrhythmogenic disorders, but not for patients with CHD [13]. The purpose of the present study was to evaluate the strategy of using ICMs for the early detection of VA in adults with CHD who are deemed at risk of VA based on their clinical profile.

2. Methods

2.1. Study design

The *Early Detection of Ventricular Arrhythmias in adults with Congenital Heart Disease using an insertable cardiac monitor* (EDVA-CHD) study is a prospective observational study which included consecutive adults

* Corresponding author at: Department of Cardiology, Erasmus MC, University Medical Center Rotterdam, dr. Molewaterplein 40, 3015 GD Rotterdam, the Netherlands.
E-mail address: s.c.yap@erasmusmc.nl (S.-C. Yap).
[1] This author takes responsibility for all aspects of the reliability and freedom from bias of the data presented and their discussed interpretation.

https://doi.org/10.1016/j.ijcard.2020.02.009

with CHD who received an ICM between March 2013 and February 2019. The starting date was chosen based on the availability of the Reveal LINQ (Medtronic Inc., Minneapolis, MN, USA) in our center. Patients who were deemed to be at risk of VA by their treating physician were eligible for an ICM. The reason for monitoring could be a combination of symptoms (e.g., (near) syncope, palpitations), prior nonsustained VA, wide QRS, and/or systemic ventricular dysfunction. The decision to implant an ICM was made in a Heart Team consisting of at least a cardiac electrophysiologist and a cardiologist specialized in CHD. The study was approved by the institutional review board of the Erasmus Medical Center. Our center is a tertiary referral center with the largest population of adults with CHD in the Netherlands.

2.2. Device programming and follow-up

All ICMs were implanted subcutaneously as recommended by the manufacturer using the incision and insertion tool. Furthermore, all patients received a handheld activator to indicate their symptoms when necessary. The ICM was usually programmed according to local settings: tachycardia-detection was set to 176 bpm for 16 beats; bradycardia-setting to 30 bpm for 8 beats; pause-setting to 4.5 s; and atrial fibrillation (AF) setting to 'AF only'. Based on the implanting physician preferences other settings could be programmed. All devices were connected to the Medtronic CareLink network for remote monitoring. Patients were discharged on the same day of implantation. Ten days after implantation the patients were seen at the out-patient clinic to check their wound and to interrogate the ICM. Afterwards, the patients were seen regularly at the outpatient clinic according to routine patient care. ICM check-ups were performed at the outpatient clinic every 6 months or earlier when necessary based on symptoms or transmitted episodes. Remote monitoring was performed on a daily basis during weekdays.

2.3. Classification of episodes and endpoints

All patient activated episodes and automatically detected episodes were classified. In the case of an inappropriate automatically detected episode, the cause of inappropriate detection was specified, if possible.

The primary endpoint of the present study was the occurrence of VA. A regular broad complex tachycardia was considered a VA if there was a sudden onset and a change in the QRS morphology in comparison to the baseline rhythm (Fig. 1A). An irregular broad complex tachycardia was considered a VA if there was a sudden onset and a polymorphic QRS morphology. A regular broad or small complex tachycardia was considered a supraventricular tachycardia (SVT) if there was a sudden onset and no change in QRS morphology (Fig. 1B). In the case of doubt, a second electrophysiologist was consulted for the final diagnosis.

The secondary endpoint was the occurrence of other arrhythmias during follow-up. Finally, it was established whether a detected arrhythmia resulted in a change in patient management ('actionable event').

2.4. Statistical analysis

Continuous data are presented as mean ± standard deviation or as median with corresponding 25th and 75th percentile, as appropriate. Categorical variables are presented by frequencies and percentages. Statistical analyses were performed using SPSS version 21.

3. Results

3.1. Study population

A total of 30 CHD patients (mean age, 38 ± 15 years; 50% male) received an ICM during the study period. Baseline characteristics of the study population are listed in Table 1. The majority of patients had moderate or severe complexity CHD. The 3 most common diagnoses were aortic coarctation, tetralogy of Fallot (TOF) and d-transposition of the great arteries (d-TGA). The majority of patients had symptoms at the time of ICM implantation (93%). An impaired systemic ventricular function was present in 17 patients (57%). A previous nonsustained VA was documented in 20% of the study population. A detailed patient-level description of CHD diagnosis, previous cardiac surgery, and reason for ICM is presented in Appendix A. There were no ICM- or procedure-related complications.

3.2. ICM-detected episodes

During a median follow-up of 16 months (interquartile range 9–-21 months), a total of 1689 episodes were transmitted to the CareLink network system (Table 2). There were 538 (32%) patient-activated episodes and 1151 (68%) automatically detected episodes. The majority of patient-activated episodes (88%) comprised sinus rhythm with or without ectopy, thus, only 12% of patient-activated episodes comprised a significant arrhythmia (Table 2).

3.3. Primary and secondary endpoints

During follow-up, 8 patients (27%) developed nonsustained VA. Four of 8 patients (50%) had a history of nonsustained VA, thus 4 patients (13%) had a de novo nonsustained VA and 4 patients (13%) had recurrent nonsustained VA. In 7 of 8 patients (88%) the VA episodes were detected by patient-activated episodes. Most patients had monomorphic VA episodes and 1 patient experienced polymorphic VA episodes. Of the 8 patients with VA, 3 patients had an impaired systemic ventricular function. Of the 8 patients with VA, 3 patients received a prophylactic ICD after consultation with their treating physician and 3 patients had a change in their antiarrhythmic drug therapy. The remaining 2 patients did not have a change in their clinical management.

A 19-year-old man with surgical corrected Shone's complex (coarctation resection, subvalvular aortic membrane resection, mitral valve and aortic valve replacement) received a dual-chamber ICD after detection of recurrent symptomatic nonsustained fast polymorphic VA (mean CL 240–270 ms, maximal 7 beats) 3 months post-ICM implantation. He received an ICM due to combination of palpitations, exercise-induced ventricular ectopy, signs of inferior wall infarction and mild impaired systemic ventricular function. After ICD implantation, he experienced two episodes of nonsustained fast polymorphic VA without ICD therapy during a follow-up of 17 months.

The second patient who received a dual-chamber ICD was a 42-year-old woman with surgical corrected TOF who experienced recurrent symptomatic nonsustained monomorphic VA (mean CL 490–520 ms, maximal 11 beats) 3 months post-ICM implantation. She received an ICM for the combination of palpitations and near-syncope. She did not experience VA post-ICD implantation during a follow-up of 27 months.

The last patient who received a dual-chamber ICD was a 44-year-old man with congenital corrected transposition of the great arteries and tricuspid valvuloplasty who developed an asymptomatic nonsustained fast monomorphic VA (mean CL 280 ms, 27 beats) 18 months post-ICM implantation (Fig. 1A). He received an ICM for a combination of syncope and dilated systemic ventricle with moderate-to-severe systolic ventricular dysfunction. During a follow-up of 12 months post-ICD implantation he experienced one episode of nonsustained fast monomorphic VA without ICD therapy.

Any significant arrhythmia was detected in 22 patients (73%). Fig. 2 shows the proportion of patients with a specific arrhythmia and Appendix A provides an overview of detected arrhythmias per patient. In 16 patients (53%) the detected arrhythmia was considered an actionable event. Management included initiation or change of antiarrhythmic drug therapy ($n = 10$, 33%), electrophysiology study ($n = 6$, 20%), ICD implantation ($n = 3$, 10%), electrical cardioversion ($n = 2$, 7%), and pacemaker implantation (n = 1, 3%).

Please cite this article as: R. Sakhi, R.M. Kauling, D.A. Theuns, et al., Early detection of ventricular arrhythmias in adults with congenital heart disease using an insertab..., International Journal of Cardiology, https://doi.org/10.1016/j.ijcard.2020.02.009

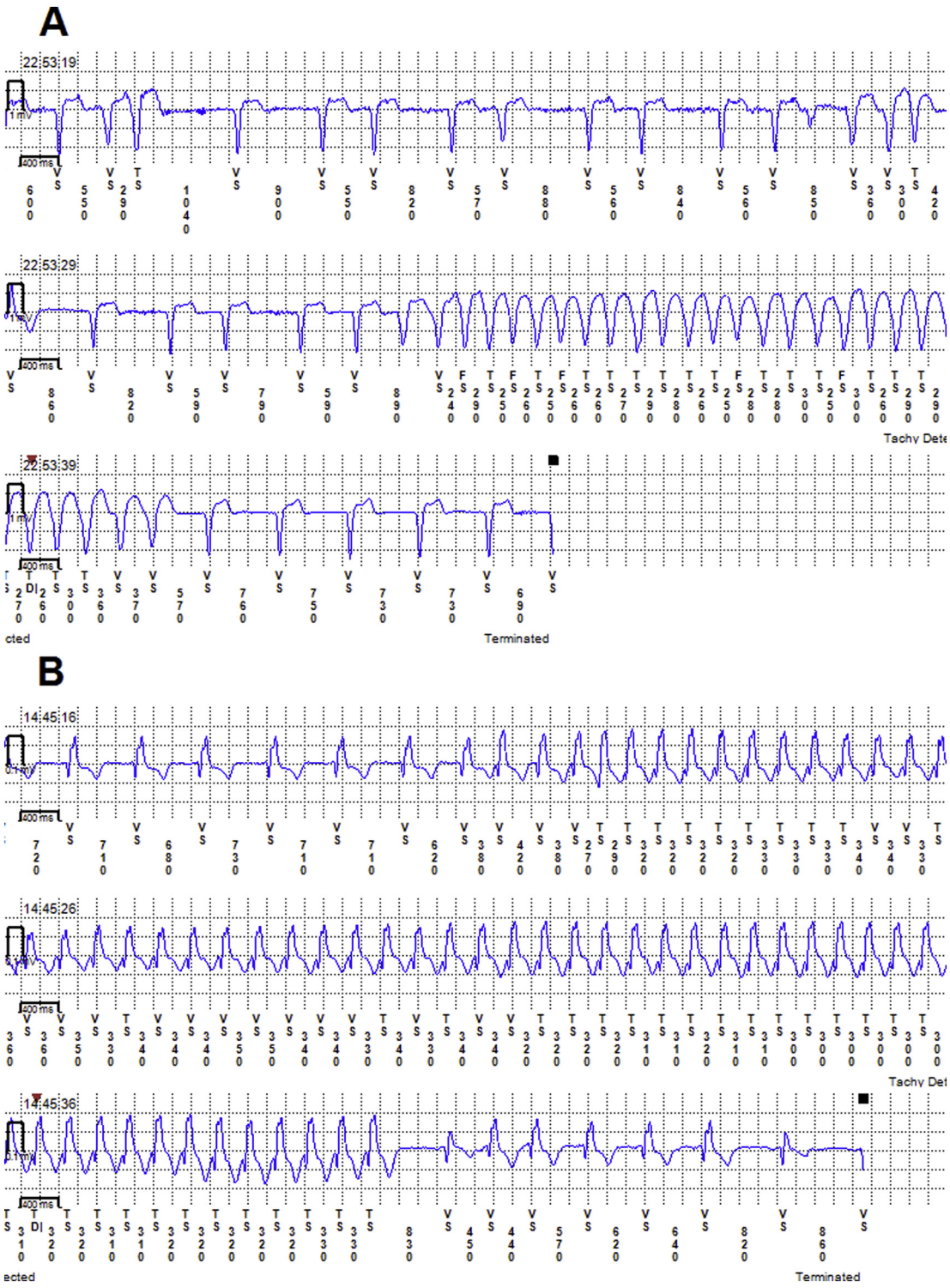

Fig. 1. Example ICM-detected episode of (A) ventricular tachycardia, (B) supraventricular tachycardia with pre-existing intraventricular conduction delay.

4. Discussion

The incidence of ICM-detected VA in a selected CHD population was relatively high (27%). The ICM-detected VA contributed to the decision to implant a prophylactic ICD in 10% of the study population. Furthermore, the detection of other arrhythmias by the ICM resulted in a significant change in clinical management in a majority of patients. The present study is the first prospective study focusing specifically on the use of an ICM for risk stratification in adults with CHD.

4.1. Risk of sudden death

Although the risk of SCD is higher in patients with CHD than in the general population, the absolute risk is still relatively low (approximately 0.1% per year) [3]. This has stimulated the search for risk factors which may help identify patients at risk for SCD, who may benefit from a prophylactic ICD implantation. Important risk factors for SCD include among others (recurrent) (non)sustained VA, inducible VA, atrial tachyarrhythmias, prolonged QRS duration, systemic ventricular dysfunction,

Please cite this article as: R. Sakhi, R.M. Kauling, D.A. Theuns, et al., Early detection of ventricular arrhythmias in adults with congenital heart disease using an insertab..., International Journal of Cardiology, https://doi.org/10.1016/j.ijcard.2020.02.009

Table 1
Baseline characteristics.

Characteristic	$N = 30$
Age, years	38 ± 15
Gender, male	15 (50%)
Hypertension	8 (27%)
Diabetes mellitus	1 (3%)
Surgical repair	25 (83%)
Symptoms:	
- Palpitations	12 (40%)
- (Near) Syncope	10 (33%)
- Palpitations and (near) syncope	6 (20%)
- Asymptomatic	2 (7%)
Congenital diagnosis:	
Aortic coarctation	7 (23%)
- AVR	3 (10%)
Tetralogy of Fallot	5 (17%)
- Transannular patch	2 (7%)
ASD	5 (17%)
- Direct surgical closure of ASD	3 (10%)
TGA corrected by atrial switch	2 (7%)
TGA corrected by arterial switch	2 (7%)
Congenital corrected TGA	2 (7%)
- Tricuspid valvuloplasty	1 (3%)
VSD	2 (7%)
- VSD patch	2 (7%)
Other	5 (17%)
Systemic systolic ventricular function:	
- Normal (EF ≥ 55%)	13 (43%)
- Mild impaired (EF 54–45%)	10 (33%)
- Moderate impaired (EF 36–44%)	7 (23%)
Electrocardiography:	
- Sinus rhythm	27 (90%)
- Other rhythm	3 (10%)
- PR interval, if sinus rhythm, ms	180 ± 49
- QRS duration, ms	136 ± 31
- QRS duration >120 ms	17 (57%)
24–48 h Holter monitoring:	
- < 1% PVCs	25 (83%)
- 1–10% PVCs	1 (3%)
- Non-sustained VA	6 (20%)
- Supraventricular tachycardia	2 (7%)
Cardiac medication:	19 (63%)
- ACE-inhibitor/ARB	8 (27%)
- Diuretics	3 (10%)
- Beta blocker	11 (37%)
- Amiodarone/Sotalol	3 (10%)
- Digoxin/calcium channel blocker	3 (10%)
- Oral anticoagulants	6 (23%)

Data is presented as n (%), mean ± SD. Abbreviations: ACE, angiotensin converting enzyme; ARB, angiotensin receptor blocker; AVR, aortic valve replacement; ASD, atrial septal defect; EF, ejection fraction; PVC, premature ventricular complex; TGA, transposition of the great arteries; VSD, ventricle septal defect; VA, ventricular arrhythmia.

Table 2
Overview ICM-detected arrhythmia episodes.

	Total ($n = 1689$)
Symptom episode:[a]	538 (32%)
Sinus rhythm	473 (88%)
- Without ectopy	314 (58%)
- With PACs	19 (4%)
- With PVCs	140 (26%)
Atrial fibrillation	33 (6%)
Regular small complex tachycardia	12 (2%)
Regular broad complex tachycardia	20 (4%)
Tachy episode:[a]	666 (39%)
Sinus rhythm	510 (77%)
- Without ectopy	268 (40%)
- With PACs	20 (3%)
- With PVCs	214 (32%)
- With noise	8 (1%)
Atrial fibrillation	8 (1%)
Regular small complex tachycardia	118 (18%)
Regular broad complex tachycardia	30 (5%)
AF episode:[a]	213 (13%)
Sinus rhythm	18 (9%)
- With PACs	2 (<1%)
- With PVCs	16 (8%)
Atrial fibrillation	180 (85%)
Small complex tachycardia	15 (7%)
- With intermittent AV-block	15 (7%)
Brady episode:[a]	147 (9%)
Sinus rhythm	41(28%)
- With undersensing due to PVCs	41(28%)
Sinus bradycardia	103 (70%)
AV block	3 (2%)
Pause episode:[a]	108 (6%)
Sinus rhythm	63 (58%)
- With sudden drop of R-wave	28 (26%)
- With small R-waves	17 (16%)
- With undersensing of PVCs	15 (14%)
- With loss of contact	3 (3%)
Sinus bradycardia	29 (27%)
AV block	3 (3%)
Sinus arrest or SA block	13 (12%)
AT episodes:[a]	17 (1%)
Atrial fibrillation	10 (59%)
Sinus tachycardia	5 (29%)
Regular small complex tachycardia	2 (12%)

Data is presented as n (%). Abbreviations: AT, atrial tachycardia; AV, atrioventricular; PAC, premature atrial complex; PVC, premature ventricular complex.
[a] Episode classification by ICM.

and subpulmonary ventricular dysfunction [4,14–17]. Despite the multitude of identified risk factors, the indication for ICD implantation remains challenging in clinical practice, especially regarding the potential downside of ICD therapy [9–11]. Furthermore, risk stratification is not uniform for the CHD population. For example, inducible sustained VA during programmed ventricular stimulation is useful for risk stratification in patients with TOF [7,8], but has not been demonstrated to predict VA/SCD in other CHD populations [18].

4.2. Risk stratification using ICM

In patients with certain high-risk features presenting with symptoms (i.e., syncope, near-syncope, palpitations), it is of importance for both patients and caregivers to rule out VA. This can be attempted using short-term monitoring; however, when symptoms are infrequent longer arrhythmia monitoring is necessary. For this purpose, an ICM is a valuable diagnostic tool for detecting paroxysmal arrhythmias as well as establishing a symptom-rhythm correlation. We provide data on the diagnostic yield of an ICM in a selected adult CHD population deemed at risk for VA. The incidence of VA was high in this population and resulted in implantation of an ICD for primary prevention in 10% of the study population. This is slightly higher than previous retrospective studies in patients with CHD who received an ICM [19,20]. These studies focused on the overall diagnostic yield of an ICM and not specifically on

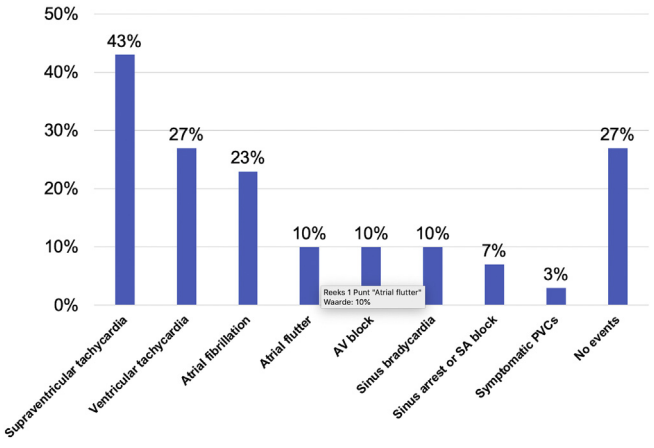

Fig. 2. Proportion of patients with an ICM-detected arrhythmia.

the role of an ICM for risk stratification. Kenny et al. described the diagnostic outcome of an ICM (Reveal or Reveal Plus, Medtronic Inc., Minneapolis, MN, USA) in a predominantly pediatric CHD population (median age 15 years) [20]. In this study, 1 of 18 patients (6%) received an ICD during a median follow-up of 19 months. The patient who received an ICD was known with Ebstein's anomaly and developed monomorphic VA at the age of 16 years. A more recent retrospective study from Boston Children's Hospital included 34 patients with CHD and an ICM (Reveal LINQ, Medtronic Inc., Minneapolis, MN, USA) [19]. In this study, 1 of 34 patients (3%) received an ICD during a median duration of follow-up of 11.8 months. The patient who received an ICD was a patient with Fontan circulation who received an ICM at the age of 32 years. Other series reporting the use of ICM in patients with CDH are smaller and mostly performed in a pediatric population [21–28].

4.3. Cost-benefit of ICMs for risk stratification

Besides the use of an ICM for risk stratification, the ICM detected a significant arrhythmia in 73% of the population and this led to a change in clinical management in 53% of patients. Therefore, an ICM can be used to titrate medication and identify candidates for an electrophysiologic procedure (i.e., pacemaker, ICD or electrophysiological study). An important aspect is the ability to differentiate between benign (near)syncope and arrhythmogenic (near)syncope. Providing reassurance to a symptomatic patient is valuable in daily clinical practice.

Although the use of ICMs for risk stratification seems promising, there are some factors which should be considered such as device costs, data overload, clinical relevance of device-detected VA and medical overuse. The issue of data overload is exemplified by the recording of >1600 episodes in 30 patients in a relatively short follow-up period in our study population. This requires a proper logistic organization with a dedicated telemonitoring staff. There is some controversy with regard to the clinical relevance of device-detected arrhythmias, especially for atrial fibrillation [12]. With regard to the clinical relevance of device-detected VA, it is important to stress that in our population the majority of VA episodes were detected as patient-activated episodes, indicating that the patient experienced symptoms. Koyak et al. previously identified that symptomatic but not asymptomatic nonsustained VA was associated with appropriate ICD therapy in TOF patients who receive an ICD for primary prevention [29].

4.4. Study limitations

A limitation of the present study is the small size and lack of a control group. Therefore, no conclusion can be made regarding the incremental value of an ICM compared to standard clinical practice with intermittent Holter monitoring. Ideally, a randomized clinical trial would be conducted where patients are randomized to an ICM or conventional follow-up. Obstacles for such a clinical trial are the heterogeneity of the population and challenges in defining appropriate endpoints. In this regard, it is important to stress that our study population was a highly selected population. The usefulness of an ICM may not apply to an unselected CHD population. Finally, the classification of broad complex tachycardia as either VA or SVT can be challenging considering that only a single surface EGM is available. To reduce the risk of misclassification, difficult EGMs were reevaluated by an electrophysiologist.

5. Conclusion

There was a high incidence of ICM-detected VA in adults with CHD who were deemed at risk of VA. ICM results led to implantation of an ICD in 10% of the study population. The detection of arrhythmias by the ICM led to important changes in the clinical management of patients. Our prospective pilot study suggests that the use of ICMs for risk stratification in selected adults with CHD is helpful.

Disclosures

Dr. Yap has received a research grant from Medtronic.

CRediT authorship contribution statement

Rafi Sakhi: Methodology, Formal analysis, Investigation, Writing - original draft, Visualization.**Robert M. Kauling:**Resources, Writing - review & editing.**Dominic A. Theuns:**Methodology, Formal analysis, Writing - review & editing.**Tamas Szili-Torok:**Conceptualization, Writing - review & editing.**Rohit E. Bhagwandien:**Investigation, Writing - review & editing.**Annemien E. van den Bosch:**Resources, Writing - review & editing.**Judith A.A.E. Cuypers:**Resources, Writing - review & editing. **Jolien W. Roos-Hesselink:**Conceptualization, Methodology, Writing - review & editing, Project administration.**Sing-Chien Yap:**Conceptualization, Methodology, Formal analysis, Writing - review & editing, Visualization, Supervision.

Declaration of competing interest

The authors report no relationships that could be construed as a conflict of interest

Appendix A

Overview of baseline characteristics and clinical outcome

	Underlying diagnosis	Surgical status	Age at repair (years)	QRS duration (ms)	Reason for ICM	Age at ICM implantation (years)	FU with ICM (months)	ICM-detected arrhythmia	ICM-guided therapy
1	Aortic coarctation	Surgical repair, AVR	8	190	Syncope, bifascicular block with first-degree AV block	56	10	Sinus arrest	AAD
2	Aortic coarctation	Surgical repair	3	117	Prior NSVT (asymptomatic)	26	12	NSVT	Conservative
3	Aortic coarctation	Surgical repair, PDA closure	11	148	Syncope, mild systemic ventricular dysfunction	71	15	SVT, AF, AFL	Ablation, ECV, AAD
4	Aortic coarctation	Surgical repair, AVR, MVR	<1	109	Palpitations, exercise-induced PVCs, mild systemic ventricular dysfunction	19	3	NSVT, SVT, SA block, AF	DDD-ICD, AAD
5	Aortic coarctation	Surgical repair, PDA closure	14	124	Palpitations, syncope, prior NSVT, mild systemic ventricular dysfunction	19	32	SVT	Ablation
6	Aortic coarctation	Surgical repair	38	81	Palpitations	41	1		
7	Aortic coarctation	Surgical repair, AVR	10	174	Palpitations, mild systemic ventricular dysfunction	55	21		

(continued on next page)

Please cite this article as: R. Sakhi, R.M. Kauling, D.A. Theuns, et al., Early detection of ventricular arrhythmias in adults with congenital heart disease using an insertab..., International Journal of Cardiology, https://doi.org/10.1016/j.ijcard.2020.02.009

(*continued*)

	Underlying diagnosis	Surgical status	Age at repair (years)	QRS duration (ms)	Reason for ICM	Age at ICM implantation (years)	FU with ICM (months)	ICM-detected arrhythmia	ICM-guided therapy
8	TOF	Total repair, PVR	30	180	Palpitations, QRS 180 ms	52	17	AF, AV block	Conservative
9	TOF	Total repair (transannular patch), PVR	<1	176	Palpitations, mild systemic ventricular dysfunction	33	17	SVT, Bradycardia	Conservative
10	TOF	Total repair (Hancock conduit)	11	155	Palpitations, near syncope	41	3	NSVT	DDD-ICD
11	TOF	Total repair	8	176	Near syncope, prior NSVT	39	21		
12	TOF	Total repair (transannular patch)	5	161	Palpitations, syncope	25	32		
13	ASD	Surgical closure ASD, PV repair	11	114	Palpitations, near syncope, mild systemic ventricular dysfunction	56	4	SVT	AAD, EPS
14	ASD	Percutaneous ASD closure	27	168	Syncope, mild systemic ventricular dysfunction	36	31	SVT, bradycardia	AAD, EPS
15	ASD			106	Syncope	74	3	AF, AV block	Pacemaker
16	ASD	Surgical closure ASD, PV repair, TV repair	8	109	Palpitations, mild systemic ventricular dysfunction	43	16	AF, SVT	Conservative
17	ASD	Surgical closure ASD, TV repair	20	116	Near syncope, mild systemic ventricular dysfunction	24	10		
18	d-TGA	Mustard repair	5	146	Near syncope, moderate systemic ventricular dysfunction, exercise-induced PVCs	39	15	AV block	Conservative
19	d-TGA	Mustard repair	4	99	Palpitations, moderate systemic ventricular dysfunction	47	4		
20	d-TGA	Arterial switch and VSD closure (patch)	<1	136	Palpitations, moderate systemic ventricular dysfunction	29	21	SVT, NSVT	Conservative
21	d-TGA	Arterial switch, VSD closure (patch)	<1	118	Palpitations, prior NSVT	18	36	SVT, NSVT	AAD
22	cc-TGA	TV repair	40	152	Syncope, prior SVT, moderate systemic ventricular dysfunction	43	18	NSVT, AF, Bradycardia	DDD-ICD
23	cc-TGA			97	Palpitations, moderate systemic ventricular dysfunction	29	30	SVT, AFL	Ablation
24	VSD	VSD closure (patch) and PDA closure	<1	104	Near syncope, prior NSVT	20	29	NSVT, SVT, AF	AAD
25	VSD	VSD closure (patch) and ASD repair	2	118	Palpitations, prior NSVT	19	12	NSVT, SVT	AAD
26	Tricuspid atresia	TCPC with lateral tunnel	11	130	Palpitations, near syncope	50	2	SVT	AAD, Ablation
27	Bicuspid aortic valve			88	Exercise-induced PVCs (asymptomatic)	41	27	Symptomatic PVCs	AAD
28	DOLV	Rastelli repair	11	174	Palpitations, prior SVT, moderate systemic ventricular dysfunction	43	12	AFL	ECV
29	Congenital PV stenosis	PV and TV repair	11	153	Palpitations, near syncope, mild systemic ventricular dysfunction	20	19		
30	Ebstein's anomaly			151	Near syncope, moderate systemic ventricular dysfunction	24	16		

Abbreviations: AAD, antiarrhythmic drug; AF, atrial fibrillation; AFL, atrial flutter; ASD, atrial septal defect; AV, atrioventricular; AVR, aortic valve replacement; cc-TGA, congenital corrected transposition of the great arteries; d-TGA, d-transposition of the great arteries; DOLV, double outlet left ventricle; ECV, electrical cardioversion; EPS, electrophysiology study; ICD, implantable cardioverter-defibrillator; ICM, insertable cardiac monitor; FU, follow-up; MVR, mitral valve replacement; NSVT, nonsustained ventricular tachycardia; PDA, patent ductus arteriosus; PV, pulmonary valve; PVC, premature ventricular complex; SA, sino-atrial; SVT, supraventricular tachycardia; TCPC, total cavopulmonary connection; TOF, tetralogy of Fallot; TV, tricuspid valve; VSD, ventricular septal defect.

References

[1] H.P. Nieminen, E.V. Jokinen, H.I. Sairanen, Causes of late deaths after pediatric cardiac surgery: a population-based study, J. Am. Coll. Cardiol. 50 (2007) 1263–1271.
[2] E.N. Oechslin, D.A. Harrison, M.S. Connelly, G.D. Webb, S.C. Siu, Mode of death in adults with congenital heart disease, Am. J. Cardiol. 86 (2000) 1111–1116.
[3] M.J. Silka, B.G. Hardy, V.D. Menashe, C.D. Morris, A population-based prospective evaluation of risk of sudden cardiac death after operation for common congenital heart defects, J. Am. Coll. Cardiol. 32 (1998) 245–251.
[4] Z. Koyak, L. Harris, J.R. de Groot, et al., Sudden cardiac death in adult congenital heart disease, Circulation 126 (2012) 1944–1954.
[5] S.C. Yap, L. Harris, Sudden cardiac death in adults with congenital heart disease, Expert. Rev. Cardiovasc. Ther. 7 (2009) 1605–1620.
[6] P. Khairy, Programmed ventricular stimulation for risk stratification in patients with tetralogy of Fallot: a Bayesian perspective, Nat Clin Pract Cardiovasc Med 4 (2007) 292–293.
[7] P. Khairy, L. Harris, M.J. Landzberg, et al., Implantable cardioverter-defibrillators in tetralogy of Fallot, Circulation 117 (2008) 363–370.
[8] P. Khairy, M.J. Landzberg, M.A. Gatzoulis, et al., Value of programmed ventricular stimulation after tetralogy of fallot repair: a multicenter study, Circulation 109 (2004) 1994–2000.
[9] S.C. Yap, J.W. Roos-Hesselink, E.S. Hoendermis, et al., Outcome of implantable cardioverter defibrillators in adults with congenital heart disease: a multi-Centre study, Eur. Heart J. 28 (2007) 1854–1861.
[10] J.T. Vehmeijer, T.F. Brouwer, J. Limpens, et al., Implantable cardioverter-defibrillators in adults with congenital heart disease: a systematic review and meta-analysis, Eur. Heart J. 37 (2016) 1439–1448.
[11] P. Opic, E.M. Utens, P. Moons, et al., Psychosocial impact of implantable cardioverter defibrillators (ICD) in young adults with tetralogy of Fallot, Clin. Res. Cardiol. 101 (2012) 509–519.
[12] R. Sakhi, D. Theuns, T. Szili-Torok, S.C. Yap, Insertable cardiac monitors: current indications and devices, Expert Rev Med Devices 16 (2019) 45–55.
13. M. Brignole, A. Moya, F.J. de Lange, et al., ESC Guidelines for the diagnosis and management of syncope, Eur. Heart. J. 39 (2018) 1883–1948.
[14] J.A. Kammeraad, C.H. van Deurzen, N. Sreeram, et al., Predictors of sudden cardiac death after mustard or Senning repair for transposition of the great arteries, J. Am. Coll. Cardiol. 44 (2004) 1095–1102.
[15] M.A. Gatzoulis, S. Balaji, S.A. Webber, et al., Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study, Lancet 356 (2000) 975–981.
[16] G.P. Diller, A. Kempny, E. Liodakis, et al., Left ventricular longitudinal function predicts life-threatening ventricular arrhythmia and death in adults with repaired tetralogy of fallot, Circulation 125 (2012) 2440–2446.

Please cite this article as: R. Sakhi, R.M. Kauling, D.A. Theuns, et al., Early detection of ventricular arrhythmias in adults with congenital heart disease using an insertab..., International Journal of Cardiology, https://doi.org/10.1016/j.ijcard.2020.02.009

[17] P. Gallego, A.E. Gonzalez, A. Sanchez-Recalde, et al., Incidence and predictors of sudden cardiac arrest in adults with congenital heart defects repaired before adult life, Am. J. Cardiol. 110 (2012) 109–117.
[18] P. Khairy, L. Harris, M.J. Landzberg, et al., Sudden death and defibrillators in transposition of the great arteries with intra-atrial baffles: a multicenter study, Circ. Arrhythm. Electrophysiol. 1 (2008) 250–257.
[19] V.J. Bezzerides, A. Walsh, M. Martuscello, et al., The real-world utility of the LINQ implantable loop recorder in pediatric and adult congenital heart patients, JACC Clin Electrophysiol 5 (2019) 245–251.
[20] D. Kenny, S. Chakrabarti, A. Ranasinghe, A. Chambers, R. Martin, G. Stuart, Single-Centre use of implantable loop recorders in patients with congenital heart disease, Europace 11 (2009) 303–307.
[21] P.A. Frangini, F. Cecchin, L. Jordao, et al., How revealing are insertable loop recorders in pediatrics? Pacing Clin. Electrophysiol. 31 (2008) 338–343.
[22] S. Sanatani, A. Peirone, C. Chiu, D.G. Human, G.J. Gross, R.M. Hamilton, Use of an implantable loop recorder in the evaluation of children with congenital heart disease, Am. Heart J. 143 (2002) 366–372.
[23] M. Gass, C. Apitz, S. Salehi-Gilani, G. Ziemer, M. Hofbeck, Use of the implantable loop recorder in children and adolescents, Cardiol. Young 16 (2006) 572–578.
[24] D.S. Kothari, F. Riddell, W. Smith, J. Voss, J.R. Skinner, Digital implantable loop recorders in the investigation of syncope in children: benefits and limitations, Heart Rhythm. 3 (2006) 1306–1312.
[25] J. Rossano, B. Bloemers, N. Sreeram, S. Balaji, M.J. Shah, Efficacy of implantable loop recorders in establishing symptom-rhythm correlation in young patients with syncope and palpitations, Pediatrics 112 (2003) e228–e233.
[26] N. Sreeram, M. Gass, C. Apitz, et al., The diagnostic yield from implantable loop recorders in children and young adults, Clin. Res. Cardiol. 97 (2008) 327–333.
[27] B. Yeung, K. McLeod, The implantable loop recorder in children, Heart 94 (2008) 888–891.
[28] A.K. Dodeja, C. Thomas, C.J. Daniels, N. Kertesz, A. Kamp, Detection of arrhythmias in adult congenital heart disease patients with LINQ(TM) implantable loop recorder, Congenit. Heart Dis. 14 (2019) 745–751.
[29] Z. Koyak, J.R. de Groot, B.J. Bouma, et al., Symptomatic but not asymptomatic non-sustained ventricular tachycardia is associated with appropriate implantable cardioverter therapy in tetralogy of Fallot, Int. J. Cardiol. 167 (2013) 1532–1535.

Please cite this article as: R. Sakhi, R.M. Kauling, D.A. Theuns, et al., Early detection of ventricular arrhythmias in adults with congenital heart disease using an insertab..., International Journal of Cardiology, https://doi.org/10.1016/j.ijcard.2020.02.009