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Conventional and right-sided screening for subcutaneous ICD in a population with congenital heart disease at high risk of sudden cardiac death

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Background: Information regarding suitability for subcutaneous defibrillator (sICD) implantation in tetralogy of Fallot (ToF) and systemic right ventricle is scarce and needs to be further explored. The main objective of our study was to determine the proportion of patients with ToF and systemic right ventricle eligible for sICD with both, standard and right-sided screening methods. Secondary objectives were: (i) to study sICD eligibility specifically in patients at high risk of sudden cardiac death, (ii) to identify independent predictors for sICD eligibility, and (iii) to compare the proportion of eligible patients in a nonselected ICD population.

Methods: We recruited 102 patients with ToF, 33 with systemic right ventricle, and 40 consecutive nonselected patients. Conventional electrocardiographic screening was performed as usual. Right-sided alternative screening was studied by positioning the left-arm and right-arm electrodes 1 cm right lateral of the xiphoid midline. The Boston Scientific ECG screening tool was utilized.

Results: In high-risk patients with ToF, eligibility was higher with right-sided screening in comparison with standard screening (61% vs. 44%; $p = .018$). Eligibility in high-risk right ventricle population was identical with both screening methods (77%, $p = \text{ns}$). The only independent predictor for sICD eligibility was QRS duration.

Conclusion: In high-risk patients with ToF, right-sided implantation of the sICD could be an alternative to a conventional ICD. In patients with a systemic right ventricle, implantation of a sICD is an alternative to a conventional sICD.

KEYWORDS

ECG screening, right-sided screening, subcutaneous implantable cardioverter-defibrillator, systemic right ventricle, tetralogy of Fallot

1 | INTRODUCTION

Sudden cardiac death (SCD) related to episodes of ventricular arrhythmias is a common cause of death in patients with either acquired or congenital heart disease. In patients with poor left ventricular systolic function, use of an implantable cardioverter-defibrillator (ICD) has been associated with a reduction in both sudden death (Køber et al., 2016) and all-cause mortality (Greenberg et al., 2004).

A high incidence of arrhythmias has been documented in patients with congenital heart disease (CHD). It is not surprising that up to 20–25% of deaths in these patients relate to SCD (Silka, Hardy, Menashe, & Morris, 1998; Oechslin, Harrison, Connelly, Webb, & Siu, 2000; Nieminen, Jokinen, & Sairanen, 2007).

Patients with tetralogy of Fallot (ToF) have a high incidence of ventricular arrhythmias and SCD (Oechslin et al., 2000). Prominent risk factors for SCD include wide QRS, prior palliative shunt, ventriculotomy,

history of nonsustained ventricular tachycardia (nSVT), and ventricular arrhythmias inducible by programmed ventricular stimulation (Khairy et al., 2010).

Patients with a subaortic right ventricle (RV), either following an atrial switch operation (Mustard or Senning) or those with congenitally corrected transposition of the great arteries (ccTGA), also have a high risk of SCD related to ventricular arrhythmias. The main predictor of SCD in this population is RV systolic dysfunction (Gallego et al., 2012; Koyak et al., 2012).

Implantation of a conventional ICD in CHD patients, who are usually young and have undergone prior surgical interventions, carries a high risk of venous occlusion, systemic embolism, endocarditis, and lead fracture (Khairy et al., 2006). There is therefore a greater need for reinterventions in these patients (Mondesert & Khairy, 2014).

In the last few years, a fully extravascular ICD has been developed (sICD) (Lambiase et al., 2014). Eliminating transvenous leads would prevent implantation-related complications (pneumothorax, cardiac tamponade), and other complications such as venous thrombosis. Also, the sICD might solve the problem of transvenous leads having limited durability. Lastly, extravascular lead placement would overcome the technical problems of difficult RV access, which arise in patients with baffle obstruction.

Unfortunately, the occurrence of inappropriate therapies has been reported, mainly in connection with T-wave oversensing phenomena (Kooiman, Knops, Olde, Wilde, & de Groot, 2014). In an attempt to mitigate this problem, an electrocardiographic screening method has been developed and tested in patient populations with acquired cardiomyopathy (dilated and ischemic) and hypertrophic cardiomyopathy (HCM). This showed over 85% eligibility for the sICD (Olde Nordkamp et al., 2014; Randles et al., 2014). There is little information as yet in adult CHD patients. Work published to date (Zeb et al., 2015; Wilson, Zeb, Veldtman, Dimitrov, & Morgan, 2016; Okamura et al., 2016) has involved small, heterogeneous samples (including patients at low, moderate, and high risk of SCD). However, sICD eligibility in CHD patients at high arrhythmic risk (systemic RV and ToF) has not yet been determined, particularly in the subgroup of individuals at greatest risk of SCD.

The main objective of our study was to determine the proportion of patients with ToF and systemic right ventricle eligible for sICD with both, standard and right-sided screening methods. Secondary objectives were: (i) to study specifically those patients at higher arrhythmic risk based on established criteria, (ii) to identify independent predictors for sICD eligibility, and (iii) to compare the results against a control group of patients in whom ICD implantation was indicated for acquired cardiomyopathy.

2 | METHODS

2.1 | Study population

One hundred and seventy-five (175) patients were recruited, divided into the following groups:

1. Adult CHD group, containing patients with the following diagnoses:

- a. Tetralogy of Fallot (ToF). Patients in this group were identified as being at significant arrhythmic risk if they scored at least 3 in the score published by Khairy et al., (2010).
 - b. Systemic right ventricle (systemic RV), due to either an atrial switch operation or ccTGA. Patients were defined as being at high arrhythmic risk if they had severely depressed RV systolic function (RVEF < 35%), as estimated by cardiac magnetic resonance imaging (CMR) (Gallego et al., 2012).
2. Control group, containing patients with a diagnosis of acquired cardiomyopathy (dilated cardiomyopathy and ischemic cardiomyopathy) in whom ICD implantation was indicated. Control patients were selected consecutively from our cardiac device clinic.

Patients were excluded if they were <18 years old or could not undergo electrocardiographic screening. Patients with a ventricular paced rhythm were not excluded, but pacing had to be programmed in bipolar mode.

Patient biometrics and clinical details were obtained. Lastly, imaging test data were collected: RV ejection fraction (RVEF) and LV ejection fraction (LVEF).

The project was approved by the institutional review board at our site, and all patients gave their informed consent to participate in the study.

2.2 | Surface electrocardiogram analysis

A resting 12-lead ECG was performed using a digital acquisition and storage system (PageWriter TC70 cardiograph, Philips, Andover, MA, USA). Specific measuring software was used for taking measurements (CardioCaliper 3.3, Iconico). Atrial and ventricular premature beats were excluded from calculations.

The following variables were obtained:

1. QRS duration (QRSd);
2. QT interval and JT interval (QT interval - QRSd), as measured in leads II, V₅ and V₆.
3. Corrected QT interval (QTc), adjusting QT duration to heart rate using Bazett's formula.

2.3 | Electrocardiographic morphology screening

Electrocardiographic screening was performed using a modified surface ECG in which the skin electrodes were positioned to represent the sICD system. For conventional ECG screening, the left-arm electrode (LA) was positioned 1 cm to the left of the sternum, at the xiphoid process, the right-arm electrode (RA) was positioned 14 cm above LA, while the left-leg electrode (LL) was positioned on the mid-axillary line, at the fifth intercostal space (Figure 1a). For right-sided parasternal ECG screening, LA and RA were moved to 1 cm right of the sternum, with the position of LL unchanged. The resulting leads I, II and III, represent the alternate, primary and secondary vectors of the sICD, respectively (Figure 1b). Ten seconds were recorded from all three leads simultaneously, using a speed of 25 mm/s and repeating

FIGURE 1 Illustration of electrode positions for conventional (a) and right-sided (b) screening. Each electrode site emulates the placement of one of the sICD sensing elements

the acquisition at three different gains (5, 10, and 20 mm/mV). Each ECG was performed supine and seated. After the acquisition of ECGs, they were analyzed using a purpose-designed tool (Figure 2). The tool determines whether R-wave amplitude is valid for detection by the sICD, and whether the R/T waves ratio is appropriate to prevent T-wave oversensing. For a patient to be eligible, at least one lead had to pass the screening test in both the supine and seated positions at the same gain.

2.4 | Statistical analysis

Data were analyzed using SPSS Statistics 18 (SPSS Inc., Chicago, IL, USA), taking statistical significance as $p < .05$.

Descriptive statistics were obtained for frequencies of continuous variables (mean $\pm$ standard deviation), as well as categorical variables (percentage $\pm$ standard deviation) (Figure 3).

Intra- and interobserver variability when measuring electrocardiographic parameters with the calipers had already been established in previous studies (Alonso et al., 2015).

The screening records obtained were analyzed in a blinded fashion by an electrophysiologist. In 10 random cases, the analysis was repeated in a blinded fashion by the same electrophysiologist and by a second observer, to determine intra- and interobserver variability.

Differences between continuous variables were examined using the Student *t* test, and categorical variables were compared using the chi-square test.

Lastly, a logistic regression model was constructed to detect independent predictors of electrocardiographic screening failure.

3 | RESULTS

3.1 | Population characteristics

The final analysis was performed on 175 patients, 102 (58%) with ToF, 33 (19%) with systemic RV, and 40 (23%) in the control group. The

clinical and electrocardiographic characteristics of the population are summarized in Table 1.

In the ToF group, 34 patients (34%) had received a prior palliative shunt and 90 (90%) had undergone ventriculotomy. Four patients (4%) showed increased LV filling pressures. Six patients (6%) had a history of nSVT on a Holter recording, and in five cases (5%) programmed ventricular stimulation was able to induce sustained VT. Lastly, 19 patients (19%) had QRSD >180 ms. Khairy score (Khairy et al., 2010) calculated for ToF patients showed a mean score of 2.4 ± 0.2 . Using a score of ≥ 3 (intermediate to high risk) as the cut-off point, 33 patients (33%) were identified as being at significant risk of SCD. The remaining 69 patients (67%) were deemed to have low SCD risk.

In the systemic RV group, 13 patients (39%) showed severely depressed RVEF ($<35\%$), and were considered at high risk of SCD (Gallego et al., 2012). Within this subgroup 12 patients had received a conventional ICD for SCD prevention.

3.2 | Intra- and interobserver variability

Evaluation of intra- and interobserver variability for electrocardiographic screening revealed extremely low variability for both conventional and right-sided screening. Both observers were consistent in all analyses performed on the selected patients, giving a kappa statistic of 1 ($p = .01$).

3.3 | Electrocardiographic screening in ToF patients

By conventional screening, 69 patients (68%) were eligible for a sICD, with a mean number of valid leads per patient of 1.23 ± 0.1 . With right-sided screening, the number of eligible patients was 77 (75%), significantly higher than with conventional screening ($p = .001$). The number of valid leads per patient was 1.33 ± 0.1 , with no significant differences seen in comparison with conventional screening ($p = .23$).

Among patients who passed conventional screening, the alternate sensing vector was the most appropriate (47 patients, 68%), followed

FIGURE 2 Illustration of the tool developed for performing conventional screening (a). The tool is placed over the ECG obtained, in the position shown in Figure 1 (c). For a complex to be considered valid, the R- or S-wave must reach the dotted line but not extend beyond the colored area, and the T-wave must stay within the colored area (b). If the T-wave extends beyond the colored area, screening is deemed to have failed (*)

by the secondary vector (39 patients, 57%). With right-sided screening, the alternate vector was again the most appropriate (43 patients, 56%), followed by the secondary vector (40 patients, 52%).

The ToF patient population was then stratified by arrhythmic risk. In the low-risk group, 55 patients (80%) passed conventional screening, and 56 (81%) passed right-sided screening ($p = ns$). In contrast, in

FIGURE 3 Example of improving in eligibility by using right-sided electrocardiographic screening. These ToF patients failed conventional screening because of a prominent T-wave (*). However, the right-sided screening smoothed ECG wave amplitude, so patients became eligible for sICD implantation (**)

the significant-risk group, sICD eligibility was significantly higher with right-sided screening (44% vs. 61%, $p = .018$).

3.4 | Electrocardiographic screening in systemic RV patients

By conventional screening, 26 patients (80%) were eligible for a sICD, with a mean number of valid leads per patient of 1.6 ± 0.18 . Right-sided screening yielded 27 eligible patients (83%), without differences to the number obtained with conventional ECG screening ($p = .5$). The number of valid leads per patient was 1.7 ± 0.2 , with no significant differences seen compared with conventional screening ($p = .6$).

In the conventional screening group, the most appropriate sensing vectors were the alternate and secondary vectors (18 patients, 70%).

With right-sided screening, the alternate vector was the most appropriate (19 patients, 72%), followed by the secondary vector (16 patients, 60%).

The systemic RV patient population was stratified according to arrhythmic risk (Gallego et al., 2012). In the high-risk group, 10 patients (77%) passed conventional and right-sided screening ($p = ns$). In the low-risk group, 16 patients (82%) passed conventional and 18 patients (88%) passed right-sided screening ($p = .24$).

3.5 | Electrocardiographic screening in pacemaker patients

Patients with an existing pacing device who had a ventricular paced rhythm during the screening were selected ($n = 10$). Results were

compared for conventional screening and right-sided screening. Higher eligibility was observed with right-sided screening (80% vs. 40%; $p = .18$). However, this was not statistically significant.

3.6 | Predictors of sICD eligibility in CHD patients

Patients who failed conventional screening showed longer QRSd (172 ± 6 vs. 131 ± 3 ms; $p < .001$) and a longer QT interval (451 ± 11 vs. 412 ± 3 ms; $p < .001$). The QTc interval was similar in both groups (483 ± 10 vs. 460 ± 9 ms; $p = .09$), as was the JT interval (287 ± 8 vs. 284 ± 3 ms; $p = .7$) (Table 2A).

Patients who failed right-sided screening showed longer QRSd (164 ± 6 vs. 114 ± 3 ms; $p < .001$) and a longer QT interval (450 ± 12 vs. 416 ± 4 ms; $p = .001$). As with conventional screening, the QTc interval was similar in both groups (488 ± 12 vs. 457 ± 8 ms; $p = .08$), as was the JT interval (284 ± 9 vs. 284 ± 3 ms; $p = .96$) (Table 2B).

The logistic regression model constructed included clinical variables such as sex, age, body mass index, type of CHD, and presence of severe RV and LV dysfunction, as well as baseline ECG data (QRSd, QT, QTc, and JT). Predictive factors for failing conventional electrocardiographic screening obtained by univariate analysis included QRSd, QT duration, and ToF. By multivariate analysis, however, the only independent predictor of conventional screening failure was QRS width (OR = 1.06, CI₉₅ 1.03–1.1; $p = .001$) (Table 3A).

The model was repeated to test for predictors of right-sided screening failure. Univariate analysis identified QRSd and QT as screening failure predictors. Multivariate analysis detected greater QRS width as an independent predictor of right-sided screening failure (OR = 1.06, CI₉₅ 1.01–1.04; $p = .004$) (Table 3B).

3.7 | Comparative eligibility of CHD patients and controls

In the control group, 37 patients (92%) were eligible for a sICD by both conventional and right-sided screening.

When these data were compared against those obtained in the ToF patient group, significant differences were apparent using conventional screening (92% vs. 68%, $p = .03$). However, these significant differences disappeared when right-sided screening was used (92% vs. 75%, $p = .1$).

When systemic RV patients were compared against the control group, no differences in sICD eligibility were seen in conventional screening (92% vs. 80% $p = .09$) or in right-sided screening (92% vs. 83%; $p = .2$).

4 | DISCUSSION

To the best of our knowledge, this is the first study to date to test sICD eligibility specifically in CHD patients at high risk of SCD.

	Control group	Tetralogy of Fallot	Systemic RV	<i>p</i>
Age (years)	61 ± 2	35 ± 1	34 ± 1	$p < .001$
Female (%)	33	37	32	$p = .85$
Weight (kg)	84 ± 4	71 ± 2	70 ± 3	$p = .009$
Height (cm)	169 ± 1	166 ± 2	169 ± 2	$p = .65$
Treatment				
Antiarrhythmic drugs <i>n</i> (%)	8 (20)	5 (5)	5 (15)	$p = .009$
Beta-blockers <i>n</i> (%)	24 (60)	30 (29)	12 (36)	$p < .001$
ARA <i>n</i> (%)	12 (30)	5 (5)	9 (27)	$p < .001$
ACEI <i>n</i> (%)	20 (50)	13 (13)	5 (15)	$p < .001$
CMR				
RVEF(%)	53 ± 4	48 ± 1	61 ± 2	$p = .001$
LVEF (%) ^a	33 ± 4	58 ± 1	43 ± 5	$p < .001$
ECG features				
QRS width (ms)	114 ± 25	150 ± 4	112 ± 10	$p < .001$
RBBB <i>n</i> (%)	4 (10)	80 (78)	9 (27)	$p < .001$
LBBB <i>n</i> (%)	4 (10)	0 (0)	2 (6)	$p < .001$
Paced QRS <i>n</i> (%)	4 (10)	5 (5)	3 (9)	$p = .4$
QT interval (ms)	400 ± 41	429 ± 5	415 ± 17	$p = .13$
JT interval (ms)	301 ± 27	282 ± 3	299 ± 6	$p = .02$
Corrected QT (ms)	450 ± 27	470 ± 9	434 ± 7	$p = .03$

TABLE 1 Baseline clinical and electrocardiographic characteristics of the study population ($n = 175$)

ARAs, aldosterone receptor antagonists; ACEI, angiotensin-converting enzyme inhibitors; CMR, cardiac magnetic resonance; RVEF, right ventricular ejection fraction; LVEF, left ventricular ejection fraction; RBBB, right bundle branch block; LBBB, complete left bundle branch block.

^aIn systemic RV patients, LVEF refers to the subaortic ventricle.

TABLE 2 Electrocardiographic characteristics in eligible patients for sICD implantation by conventional screening (A) and right-sided screening (B)

	Passes conventional screening	Fails conventional screening	<i>p</i>
(A)			
QRS width (ms)	131 ± 3	172 ± 6	<i>p</i> < .001
QT interval (ms)	412 ± 3	451 ± 11	<i>p</i> < .001
Corrected QT (ms)	460 ± 9	483 ± 10	<i>p</i> = .09
JT interval (ms)	284 ± 3	287 ± 8	<i>p</i> = .96
	Passes right-sided screening	Fails right-sided screening	<i>p</i>
(B)			
QRS width (ms)	114 ± 3	164 ± 6	<i>p</i> = .002
QT interval (ms)	416 ± 4	450 ± 12	<i>p</i> = .001
Corrected QT (ms)	457 ± 8	488 ± 12	<i>p</i> = .08
JT interval (ms)	284 ± 9	284 ± 3	<i>p</i> = .96

TABLE 3 (A) Logistic regression model to predict conventional screening failure (B) Logistic regression model to predict right-parasternal screening failure

	Univariate analysis		Multivariate analysis	
	OR	<i>p</i>	OR	<i>p</i>
(A)				
Age (years)	3.6	.06		
Female	1.01	.32		
Weight (kg)	0.03	.87		
Height (cm)	1.3	.26		
ToF	3.9	.048		
RVEF <35%	0.3	.86		
LVEF <35%	0.13	.7		
QRSd (ms)	26.9	.001	1.06	.001
QT (ms)	18.6	.001		
JT (ms)	0.31	.6		
QTc (ms)	2.1	.15		
	Univariate analysis		Multivariate analysis	
	OR	<i>p</i>	OR	<i>p</i>
(B)				
Age (years)	1.16	.23		
Female	0.44	.51		
Weight (kg)	0.05	.83		
Height (cm)	0.05	.95		
ToF	2.17	.14		
RVEF <35%	0.08	.77		
LVEF <35%	0.35	.55		
QRSd (ms)	8.69	.002	1.03	.004
QT (ms)	9.73	.003		
JT (ms)	0.56	.45		
QTc (ms)	1.61	.2		

The main findings of this study were: (i) 44% of ToF patients with significant arrhythmic risk were eligible for a sICD by conventional screening; (ii) using right-sided screening achieved a significant increase in sICD

eligibility in ToF patients with significant arrhythmic risk; (iii) sICD eligibility in ToF patients was lower than in the control group; and (iv) patients with systemic RV showed similar sICD eligibility as the control group.

ToF patients with arrhythmic risk factors (Khairy et al., 2010) may benefit from ICD implantation as primary prevention of SCD. Likewise, in systemic RV patients with severe systolic dysfunction, ICD implantation is recommended as primary prevention of SCD (Gallego et al., 2012).

Young patients have a high risk of defibrillation lead complications, because they are more physically active and usually live longer. They are also susceptible to technical difficulties caused by venous system obstruction in patients with repaired CHD (Khairy et al., 2006; Mondesert & Khairy, 2014). Implanting a sICD eliminates the need for a transvenous lead, so offers an interesting alternative to a conventional ICD in this patient group.

Preimplantation electrocardiographic screening is required to identify patients eligible for a sICD. Screening was previously studied in patients with acquired cardiomyopathy, and showed 88% eligibility (Randles et al., 2014). Francia et al. examined ECG screening in patients with hypertrophic cardiomyopathy, obtaining 90% sICD eligibility, but fewer valid vectors (Alonso et al., 2017). In the CHD context, Zeb et al. tested sICD eligibility in three groups of patients diagnosed with CHD (systemic RV, ToF, and Fontan physiology). In all, the study included 30 patients (Zeb et al., 2015). Zeb concluded that eligibility in CHD patients was similar to that of patients with acquired cardiomyopathy. A recent study (Okamura et al., 2016) analyzed the role of right-sided parasternal screening in nonselected CHD patients (22 ToF and seven systemic RV), concluding that eligibility in CHD patients is lower than in the general population, but might be equivalent using right-sided screening. More recently, our group published the initial experience with ToF patients; we showed an increase in sICD eligibility in ToF patients when right-sided screening was utilized (Alonso et al., 2017). Altogether, studies published to date have involved 92 patients with ToF and 17 with systemic RV. These patients were not selected, and we do not know the details of their systolic function or other markers for SCD risk. Our study aimed to test sICD eligibility in a large sample of CHD patients, stratified by estimated SCD risk.

4.1 | Patients with tetralogy of Fallot

Eligibility in the general ToF patient population was lower than in the control group using conventional screening (68% vs. 92%, $p = .03$), and showed no differences using right-sided screening (75% vs. 92%, $p = .1$). Discrepancies from published results are probably due to the characteristics of our population, which included older patients with a high prevalence of pulmonary regurgitation, resulting in higher indexed RV end-diastolic volumes and greater QRS width. The only parameter that independently predicted screening failure was precisely QRS width, which probably explains the low sICD eligibility in our study.

The effect of QRS width (QRSd > 180 ms) in patients with ToF is well known, and has been related to SCD (Khairy et al., 2010; Gatzoulis et al., 2000). Nonetheless, QRSd is not the only risk factor for SCD in ToF patients. Other factors, as LV dysfunction, have been strongly associated with ventricular arrhythmias and SCD in this population (Diller et al., 2012). Our work showed that patients with prolonged QRS were less eligible for sICD implantation and QRSd was the only

independent predictor of screening failure. Thus, in patients with high SCD risk and a wide QRS complex sICD eligibility should be carefully assessed with conventional and right-sided ECG screening. In these patients ECG screening during exercise test might be recommended in order to avoid false positive results.

Our study is the first to show that eligibility by conventional screening in ToF patients with significant SCD risk is as low as 44%. Eligibility in this patient subgroup increased by 39% with right-sided screening, so implanting a sICD with the defibrillation lead in a right parasternal position could be a feasible option in these patients. One possible explanation for this phenomenon is attenuation of the effect of right ventricular hypertrophy by the interposed sternum and the longer sensing vector.

4.2 | Patients with systemic RV

Systemic RV patients showed eligibility similar to the control group, with both conventional and right-sided screening. This eligibility was unchanged when patients at greater arrhythmic risk were selected from this population, being consistent with results reported previously in non-selected patients (Zeb et al., 2015). Implanting a sICD would be a possible option in systemic RV patients in whom a defibrillator is indicated.

It is not uncommon for systemic RV patients to develop complete heart block requiring permanent ventricular pacing, which might represent a limitation for sICD therapy. In our study, in the pacemaker subgroup, eligibility by right-sided screening was similar to that obtained in the general population (80%), whereas eligibility by conventional screening was low (40%). These data suggest that, in patients with an existing pacemaker, right-sided parasternal sICD implantation could be an alternative to transvenous ICD implantation. Also, recent data support the concomitant use of a sICD and a permanent pacemaker (Huang, Patton, & Prutkin, 2016).

In contrast to data obtained in the general population (Olde Nordkamp et al., 2014) the alternate vector was the one that proved valid most often, in both ToF and systemic RV patients. This might be explained by the alternate vector being perpendicular to the longitudinal axis of the heart, producing an electrical signal of smaller amplitude, and minimizing the risk of T-wave oversensing. These findings are consistent with those seen in patients with hypertrophic cardiomyopathy (Francia et al., 2015).

4.3 | Limitations

Our study has several limitations. Firstly, it was a single-center study with a relatively small sample size, which limits its statistical power. However, given the low prevalence of the pathologies considered in this work, we think the sample size is acceptable.

Secondly, all screening tests were performed at rest (seated and standing), so no conclusions can be drawn about what might happen during exercise.

Thirdly, the criterion used to define patients at significant risk in the ToF population was the Khairy score (Khairy et al., 2010). Although the use of these criteria is controversial, no highly specific criteria for

predicting SCD currently exist. What is evident is that QRS width (mainly QRSd > 180 ms) is highly predictive of SCD in work published to date (Khairy et al., 2010; Gatzoulis et al., 2000).

Finally, the number of patients with a permanent pacemaker included in the study was low; thus, the conclusions should be taken with caution.

5 | CONCLUSIONS

Our study is the first to show that, in ToF patients at significant risk of SCD, sICD eligibility is low by conventional screening, but increases significantly when right-sided screening is used. Therefore, right-sided sICD implantation is a feasible option in many ToF patients in whom an implantable defibrillator is indicated.

Patients with systemic RV in whom implantation of a defibrillator was indicated showed similar eligibility as the general population, so a sICD is a possible option in this population, even in patients with an existing pacemaker.

Noneligible patients had a wider QRS complex and a longer QT interval. In addition, QRS duration was identified as the only independent predictor of sICD screening failure.

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