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REVIEW

Idiopathic short-coupled ventricular tachyarrhythmias: Systematic review and validation of electrocardiographic indices

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ABSTRACT

Introduction: Idiopathic short-coupled ventricular tachyarrhythmias make up a considerable proportion of ventricular tachyarrhythmias in structurally normal hearts and are the cause of 5–10% of unexpected sudden cardiac deaths. There is disparity in the literature regarding their description and a lack of formal diagnostic criteria to define them.

Objective: To validate ECG indices for the diagnosis of these ventricular tachyarrhythmias and to subsequently unify their differing descriptions in the literature under a new terminology: *Idiopathic Short-Coupled Ventricular Tachyarrhythmias*.

Methods: We conducted a systematic review of all published studies describing short-coupled torsades de pointes, idiopathic ventricular fibrillation and polymorphic ventricular tachycardia. Published tracings were analysed using a standard set of criteria to define the different ECG intervals. Previously proposed diagnostic indices were validated using a control group of previously published long-coupled torsades de pointes cases.

Results: Validation of the ECG indices revealed that a coupling interval < 400 ms was the most reliable measurement (sensitivity 100%, specificity 97%), followed by a coupling interval/QT < 1 (sensitivity 96%, specificity 100%).

Conclusion: Idiopathic short-coupled ventricular tachyarrhythmias encompass all previous descriptions of this tachyarrhythmia including idiopathic ventricular fibrillation, short-coupled torsades de pointes, Purkinje-related torsades de pointes and idiopathic polymorphic ventricular tachycardia. This arrhythmia can be diagnosed by newly proposed criteria with high sensitivity and specificity.

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Abbreviations: ECG, electrocardiography; CI, coupling interval; CA, cardiac arrest; ISCVT, idiopathic short-coupled ventricular tachyarrhythmia; IVF, idiopathic ventricular fibrillation; SCTDP, short-coupled torsades de pointes; SCD, sudden cardiac death; LCTDP, long-coupled torsades de pointes; MESH, medical subject headings; PMT, polymorphous ventricular tachycardia; Pr-TDP, Purkinje related torsades de pointes; Pal/Syn, palpitations/syncope; PVC, Premature Ventricular Contraction; RVOT, right ventricular outflow tachycardia; TDP, torsades de pointes; T_{asc}, ascending limb of the T wave; T_{desc}, descending limb of the T wave; VF, ventricular fibrillation.

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1. Introduction

Idiopathic ventricular tachyarrhythmias represent a significant proportion (5–10%) of all ventricular arrhythmias.¹ A growing body of evidence suggests that Purkinje fibres play a pivotal role in these idiopathic forms,^{2–5} but there is no agreement on the mechanism of this rare type. In the last two decades, different eponyms of these arrhythmias have been used which may or may not include suggestions as to the underlying mechanisms involved in their generation. These include idiopathic ventricular fibrillation³ (IVF), Purkinje-related torsades de pointes⁵ (Pr-TDP), short-coupled torsades de pointes⁵ (SCTDP) and polymorphous ventricular tachycardia⁷ (PMT) in structurally normal heart. These different terms may indeed be referring to the same entity.

We systematically reviewed the literature to identify reports of these arrhythmias, validate ECG algorithms for their diagnosis, and propose a unifying terminology.

2. Material and methods

We searched Medline and Pre-Medline (PubMed) and Embase (Ovid) versions from 1992 to present (last update December 2015) to identify all observational studies, review articles and case reports describing idiopathic forms of ventricular arrhythmia. Mesh terms used in the search were: “ventricular arrhythmia”, “short coupled”, “torsades de pointes”. After our initial search identified several reports describing short-coupled torsades de pointes but labelling them as PMT or IVF, we expanded our search to include the term IVF. We further reviewed reference lists within articles to complement our electronic searches.

Our main objective was to identify ECG features that may be common to the different cases of IVF described in the literature. To serve as a control to the ECG features of IVF, we also performed a limited search using PubMed and included 12 case reports each of acquired and congenital LQT syndrome with TDP episodes. The purpose of the latter was to validate previously published ECG indices used to discern “what we described as ISCVT in this study” from torsade de points that shares similar polymorphic pattern. We restricted the search to papers published in English. The search strategies are available on request.

2.1. Study selection

We selected articles using two reviewers (MA, AA). The full text manuscripts and the extracted data were reviewed carefully to ensure that all discussing the same arrhythmia. The quality of the published ECGs and/or rhythm strips was also reviewed to ensure accurate measurements of the intervals. Disagreement was solved by consensus. Published 12 lead ECGs or rhythm strips were reviewed and relevant intervals were measured by one reviewer using an electronic calliper, blinded to the study's published measurements (Figs. 1 and 2). ECGs or rhythm strips without clear scale standards or a well-defined QT interval of a normally conducted ventricular beat were excluded from the analysis.

2.2. Data extraction

Data were extracted in duplicate, including the number and characteristics of patients (age, gender, presence of structural heart disease or other co-morbidities). History of associated syncope or resuscitated cardiac arrest was also extracted. The calculation of the electrocardiographic indices was performed on the published rhythm strips/ECGs in each article. All extracted and calculated indices were plotted in a constructed two by two contingency table. Indices from the acquired and congenital Long QT tachyarrhythmia were used as a control group to validate ECG diagnostic indices of idiopathic forms (Table 2).

2.3. ECG parameters and definitions

The following parameters were measured:

1. Coupling interval (CI): CI was defined as the interval measured from the initial deflection of the preceding QRS to the initial deflection of the coupled premature ventricular complex (PVC), Fig. 1.
2. CI/QT ratio was defined as the ratio of the PVC CI and the QT interval of the preceding normally conducted ventricular beat, Fig. 1. Tangent angle technique was utilised to determine the exact terminal point of the T wave. No standard ECG lead was

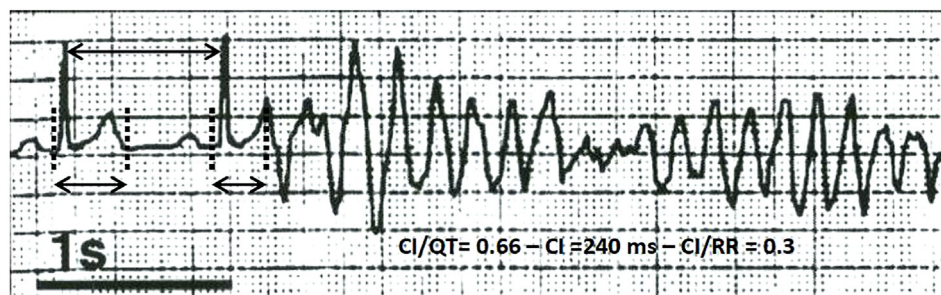


Fig. 1. ISCVT episode with calculation method of electrocardiographic indices. Shiga et al.³⁵, ISCVT = idiopathic short coupled ventricular tachyarrhythmia.

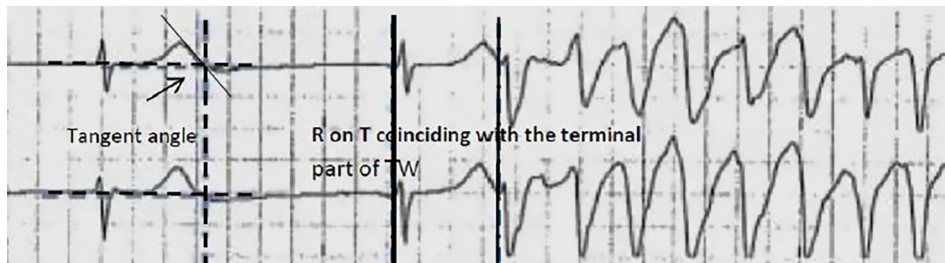


Fig. 2. Example of LCTDP episode demonstrating coinciding R on terminal part of the TW. LCTDP = long coupled torsade de pointes, TW = T wave.

specifically used to perform the measurements due to the variation in the published ECGs/tracings. If more than one lead was available for measurement, we used the one with clearest tracing. A CI/QT ratio of <1 was used to define “short-coupling” PVC.⁷

2.4. Statistical analysis

Data were entered into an Excel spreadsheet and imported into IBM SPSS (Version 21.0 for Windows, Armonk, New York) for statistical analysis. Standard descriptive statistics were used to summarize continuous data with means/standard deviation and categorical data with frequencies and percentages. Optimal cut-points for CI/QT and CI, for both the studied cohort and the control group of LQT, were identified using ROC Curves. Sensitivity and specificity for the CI/QT ratio and CI thresholds were calculated. Chi-Square test was used to calculate the P value of 2×2 contingency data.

3. Results

Our search strategy identified 42 articles that were further reviewed for reported cases of IVF, SCTDP, PMT and Pr-TDP. A total of 32 articles described single cases with the inclusion of relevant clinical details and measurable intervals on the surface ECG. The other 10 articles were excluded due to poor quality of the published ECGs or rhythm strips. A detailed listing of the clinical and electrocardiographic characteristics of the studied cohort is shown in Table 1. Most patients were male ($n = 17$), and approximately half of them were 40 years of age or younger ($n = 17$). All patients except two^{8,9} had documented normal cardiac structure by echocardiography. In the two papers^{8,9} where the authors did not discuss work up details, they settled for describing the arrhythmia as being “idiopathic”. In accordance to that we assumed that there was no evident structural heart disease in those subjects and therefore were considered in the study. Family history of SCD was documented in only one patient. No single case was described

Table 1
Patients characteristic, electrocardiographic findings and calculated indices.

Article	Sex	Age	Presentation	BBB	PVC morph	Episode	Pause	CI/QT	CI/RR	CI	R on T
Leenhardt et al. ⁶ SCTDP	F	31	Pre-syncope/Syncope	LBBB	POLYM	PMT/VF	NO	0.67	0.25	255	Asc limb
Leenhardt et al. ⁶ SCTDP	M	15	Pre-syncope/Syncope	RBBB	UNI	PMT/VF	NO	0.7	0.35	280	Asc limb
Eisenberg et al. ⁷ PMT	–	36	Pre-syncope/Syncope	–	UNI	PMT	YES	1	0.33	408	Desc limb
Eisenberg et al. ⁷ PMT	F	58	Cardiac Arrest	–	–	PMT	NO	0.85	0.5	355	Desc limb
Anter E. et al. ¹¹ IVF	M	59	Cardiac Arrest	LBBB	UNI	PMT/VF	NO	0.65	0.2	290	Asc limb
Takeuchi T et al. ¹² SCTDP	M	51	Cardiac Arrest	RBBB	UNI	PMT/VF	YES	0.7	0.27	260	Desc limb
Viskin et al. ⁸ IVF	F	50	Cardiac Arrest	LBBB	POLYM	PMT/VF	NO	0.75	0.4	300	Asc limb
Viskin et al. ¹³ SCTDP	F	55	Pre-syncope/Syncope	LBBB	UNI	PMT	NO	0.89	0.3	330	Desc limb
Suh et al. ¹⁷ IVF	F	40	Cardiac Arrest	LBBB	UNI	PMT	NO	0.75	0.47	270	Asc limb
Betts et al. ¹⁸ IVF	M	32	Cardiac Arrest	LBBB	UNI	PMT	NO	0.7	0.3	300	Asc limb
Allocca et al. ¹⁹ IVF	F	47	Cardiac Arrest	LBBB	–	PMT	NO	0.87	0.58	365	Desc limb
Mechleb et al. ⁹ PMT	F	55	Cardiac Arrest	LBBB	–	PMT	NO	0.86	0.42	355	Desc limb
Burrows et al. ²⁰ SCTDP	F	24	Cardiac Arrest	LBBB	UNI	PMT	NO	0.85	0.41	330	Desc limb
Mounz et al. ²¹ SCTDP	–	–	–	–	UNI	PMT	NO	0.54	0.27	200	Asc limb
Nogami et al. ²² IVF	M	54	Cardiac Arrest	RBBB	UNI	PMT	NO	0.81	0.48	260	Asc limb
Brendan et al. ²³ SCTDP	M	51	Cardiac Arrest	LBBB	–	PMT	NO	0.71	0.39	270	Asc limb
Chiladakis et al. ²⁴ SCTDP	F	50	Cardiac Arrest	–	–	PMT/VF	NO	0.73	0.3	345	Asc limb
Mourad et al. ²⁵ SCTDP	M	38	Cardiac Arrest	LBBB	POLYM	PMT	NO	0.85	0.36	325	Desc limb
Almehairi et al.	M	25	Cardiac Arrest	RBBB	UNI	PMT	NO	0.83	0.52	275	Desc limb
Landen et al. ²⁶ PMT	M	53	Cardiac Arrest	LBBB	UNI	PMT	NO	0.79	0.59	346	Asc limb
Rak Cho et al. ²⁷ IVF	M	17	Cardiac Arrest	LBBB	UNI	PMT	YES	0.92	0.26	370	Desc limb
Aizawa Y et al. ²⁸ IVF	M	13	Cardiac arrest	RBBB	UNI	PMT	YES	0.9	0.45	370	Desc limb
Takatsuki et al. ²⁹ IVF	M	62	Pre-syncope/Syncope	LBBB	UNI	PMT	YES	–	0.3	320	Desc limb
Naik N et al. ³⁰ IVF	M	24	Pre-syncope/Syncope	LBBB	UNI	PMT	NO	0.8	0.34	300	Desc limb
Kohsaka S et al. ³¹ IVF	F	21	Pre-syncope/Syncope	LBBB	UNI	PMT	NO	0.8	0.41	300	Desc limb
Yamazaki M et al. ³² SCTDP	M	21	Pre-syncope/Syncope	LBBB	UNI	PMT	NO	0.84	0.28	350	Desc limb
Shiga et al. ³³ SCTDP	M	41	Pre-syncope/Syncope	LBBB	UNI	PMT/VF	NO	0.66	0.3	240	Asc limb
Wafa et al. ³⁴ SCTDP	F	30	Pre-syncope/Syncope	–	UNI	PMT	YES	0.78	0.27	300	Desc limb
Yanfei et al. ³⁵ SCTDP	M	30–40	Pre-syncope/Syncope	–	UNI	PMT	NO	0.7	–	280	Asc limb
Yanfei et al. ³⁵ SCTDP	F	30–40	Pre-syncope/Syncope	–	UNI	PMT	–	0.65	–	230	Desc limb
Yanfei et al. ³⁵ SCTDP	F	30–40	Pre-syncope/Syncope	–	UNI	PMT	–	0.71	–	270	Peak
Bogaard et al. ³⁶ SCTDP	M	36	Pre-syncope/Syncope	LBBB	UNI	PMT	NO	0.78	0.34	300	Asc limb

Table 2
Calculated indices for the control group of acquired and congenital long QT syndrome.

Congenital	QT/RR Index	CI	CI/QT index
Sami et al. ¹⁵	0.41 ms	640 ms	1.04 ms
Monahan et al. ³⁷	0.56 ms	707 ms	1.06 ms
Sami et al. ³⁸	0.57 ms	600 ms	1.03 ms
Shimizu ³⁹	0.35 ms	630 ms	1.30 ms
Basamad et al. ⁴⁰	0.36 ms	765 ms	1.25 ms
Figuroa et al. ⁴¹	0.44 ms	640 ms	1.25 ms
Acquired	QT/RR index	CI	CI/QT index
Gregorio et al. ⁴²	0.52 ms	590 ms	1.00 ms
Gibson et al. ⁴³	0.3 ms	635 ms	1.35 ms
Hiede et al. ⁴⁴	0.4 ms	495 ms	1.21 ms
Arce et al. ⁴⁵	0.4 ms	588 ms	1.06 ms
Colombo et al. ⁴⁶	0.58 ms	640 ms	1.2 ms
Bass et al. ⁴⁷	0.3 ms	400 ms	1.05 ms

in association with exercise. All cases were reported to have normal electrolyte and metabolic indices. Only one article was published as a pure ECG case of SCTDP, with no further clinical data.

Work up for evidence of coronary artery disease (CAD) was described in 31 cases. No obstructive coronary artery disease was reported in the 27 cases in whom coronary angiograms were performed. In the other three cases CAD was ruled out by non-invasive methods. Cardiac MRI was only performed in three patients and showed normal cardiac structure. RV biopsy was performed in 3 patients and showed no evidence of arrhythmogenic right ventricular dysplasia.

Successful ablation for PVCs foci was achieved in 7 subjects. One subject underwent electrophysiology study and was negative for inducible arrhythmia following verapamil therapy.

Genetic testing was only done in one subject and was negative for inherited channelopathies.

Fourteen subjects received ICD implantation whereas 13 subjects were maintained on verapamil and 4 on B blockers (atenolol and propranolol).

3.1. Clinical features

The age was reported in 32 subjects only and the mean was calculated as ($n = 39.9 \pm 13.7$) and the median as 40 years. Gender was reported in 26 subjects of which 14 were males and 12 were females.

Reported symptoms were aborted cardiac arrest (CA) in 17/32 and palpitations or syncope in 14/32. One case was reported without description of symptoms and only included ECG findings (Table 1).

3.2. ECG characteristics

There were 27 cases with identifiable ectopic beats that underwent further analysis for the purposes of consistency of which 24 presented unifocal morphology.

Pauses preceding the short-coupled arrhythmia was identified in 5/28 subjects. Four patients in this category presented clinically with palpitation/syncope whereas only one presented with CA.

Pure polymorphic pattern was noticed in 24 patients of the total cohort and 7 patients degenerated into VF.

3.3. Validation of the ECG indices

The CI/QT index was applied with a score of < 1 used to specify the short-coupled arrhythmia. CI/QT index of < 1 was 96% sensitive and 100% specific for it. The mean CI for the short coupled arrhythmia and LCTDP were 304 ± 47 ms and 638.08 ± 47.177 ms; respectively (95% CI, 294.6–379.7). Two cut-off points for CIs were

Table 3
Sensitivity and specificity analysis of the electrocardiographic indices.

ECG indices	Se & Sp
CI \leq 300 ms	100–50%
CI $<$ 400 ms	100–97%
CI/QT $<$ 1 ms	96–100%

analysed for their accuracy in identifying the short-coupled arrhythmia. A CI $<$ 400 ms was 100% sensitive 97% specific, whereas CI \leq 300 ms is 100% sensitive and 50% specific (Table 3).

3.4. Timing of coinciding PVCs with the T waves

Comparative analysis of the coinciding PVCs on the ascending (T_{asc}) and descending (T_{desc}) limbs of the T wave were performed to assess the predictability of the clinical presentation. R on T_{asc} was observed in a total of 14 patients, 64% in the CA group and 36% in the Pal/Syn group. R on T_{desc} was comparatively seen in a total of 18 subjects, 44% in CA group and 55% in Pal/Syn group. The correlation between the type of clinical presentation and the timing of R on T was not statistically significant (P value 0.47). Interestingly, 6 patients from 7 who presented with VF degeneration had R on T_{asc} . Despite that, CA occurred in some of them.

3.5. Relation of the coupled PVCs to the Purkinje system

Of the 9 patients that underwent further ectopic mapping and ablation, 5 were identified to be Purkinje related arrhythmia whereas the remaining 4 were found to be myocardial in origin.

4. Discussion

This type of short-coupled arrhythmias is rare and accounts for 5–10% of out-of hospital aborted cardiac arrests.¹ Variable eponyms used to describe this arrhythmia created confusion surrounding its underlying mechanism. We reviewed the literature for case reports/series describing IVF, SCTDP, Pr-TDP and idiopathic PMT and the similarities in their features indicated that all presenting same type of arrhythmia.

Leenhardt et al.⁶ first described an arrhythmia they termed SCTDP in 14 patients with structurally normal hearts without associated channelopathies. Yet the term TDP historically relates to long QT arrhythmias that conceptually conflicts with the proposed mechanism for a TDP specified as short-coupled. In contrast, Eisenberg et al.⁷ utilized the term polymorphous VT in structurally normal heart, a description that is not sensitive enough to exclude inherited channelopathies that could complicate similar pattern of short-coupled arrhythmias. Haïssaguerre et al.³ and Knecht et al.¹⁰ preferred the descriptive term IVF, whereas Nogami⁵ refined Leenhardt's description to be Pr-TDP.

One may argue that using IVF as a unifying diagnostic term potentially creates conceptual conflicts for an arrhythmia that is always PMT/TDP in pattern and combined with degeneration to VF in some cases^{11,12}. VF degeneration, duration of the arrhythmia, frequency of recurrences/shocks and clinical presentation, could have a close relation to the coinciding PVCs on the T-wave. Viskin et al.⁸ indicated that the propensity for poor outcomes associates with presentations of coinciding R on T_{asc} . Alshekh-Ali et al.¹⁴ demonstrated that VF induction in an experimental commotio cordis model has close relationship with critical timing of chest impact during the repolarization phase. As indicated above, we deemed CA type of initial clinical presentation a poor outcome. An inference from the coinciding PVC on the T wave (ascending

or descending) was attempted to assess the propensity for the type of the clinical presentation. The comparative analysis suggested that R on T_{asc} or T_{desc} does not predict the likelihood of CA over Pal/Syn type of presentation, ($P=0.47$). However, 7 patients in from the total cohort presented with true VF degeneration and 6 had a significant correlation with R on T_{asc}, which may indicate that the R on T timing plays a significant role in VF degeneration but does not necessarily lead to CA. Notably, fifteen patients (45%) presented with syncope of which 3 had occasional VF degeneration during the episode. Leenhardt's group has described SCTDP purely consisting of PMT/TDP pattern, while in some cases has been in association with VF degeneration. In contrast, the majority of IVF cases presented as pure PMT/TDP pattern, although this observation was limited by the available ECGs displayed in the studied articles. Nogami et al.⁵ specified SCTDP as Purkinje-related arrhythmia and used this terminology interchangeably between the two entities. In the studied cohort, however, 4 of 9 patients underwent formal Electrophysiological Study and were identified to have non-Purkinje related PVCs. Similarly, Both Haïssaguerre et al.³ and Knecht et al.¹⁰ reported non-Purkinje related PVCs in some cases.

We propose the term “*idiopathic short-coupled ventricular tachycarrhythmias*” to be a unifying eponym for this group of ventricular tachycarrhythmias. “*Idiopathic*” is proposed to avoid an overlap with aetiologies known to precipitate similar short-coupled ventricular arrhythmias such as catecholaminergic polymorphic ventricular tachycardia, Brugada syndrome, short QT syndrome or ischemia. “*Short-coupled*” characterises those critically coupled PVCs which are required to occur in the vulnerable phase, in the context of normal cardiac structure. *Ventricular Tachycarrhythmias* was used as a general term to accommodate any possible form of this arrhythmia, whether it is polymorphic VT, VF or in both combinations. As known, pauses almost always are the trigger in LCTDP^{15,16}; however, our search revealed 5 patients who demonstrated pauses preceding the short-coupled arrhythmia and yet had no association with long QT syndrome. Validation of the CI that was depicted by Leenhardt et al.⁶ as ≤ 300 ms was performed. The purpose was to set a discriminating threshold to help rule out LCTDP. In that study, 14 patients were studied and the longest CI was 300 ms. We applied this criterion to our cohort and found a sensitivity of 100%, but with a specificity that did not exceed 50%. Of the several CIs analysed, the most effective threshold was found to be < 400 ms, which demonstrated 100% sensitivity and 97 % specificity. Eisenberg et al.⁷ has proposed the CI/QT index as a useful discriminator of idiopathic subtypes of PMT in structurally normal hearts. Our validation of this index is in agreement, with sensitivity and specificity of 96% and 100%, respectively. Several case reports and two original studies^{3,10} showed that ablation strategy in these cases is of great clinical benefit. Our analysis revealed that 9 patients underwent successful suppression of PVCs with radiofrequency ablation. Knecht et al.¹⁰ reported successful ablation of 38 patients in a multi-centre trial with 89% of patients remaining free of recurrences at 5-year follow up. Additionally, Haïssaguerre et al.³ achieved successful ectopics ablation in 27 patients. It is important to note that attempting ablation is a purely palliative strategy and that defibrillator implantation is mandatory to prevent SCD.³ Conservative medical therapy using verapamil has been shown in a majority of patients to be a valuable strategy in suppressing PVCs, reducing the frequency of the short-coupled arrhythmia and delaying the CI of the PVC.^{6,7}

5. Limitations

We present a systematic review of case reports published under different and unrelated titles. Many ECGs and tracings met only

moderate quality in terms of their clarity that necessitated computer based editing. Many case reports used Leenhardt's term SCTDP or PMT, which precluded them from further invasive investigation to determine if they were related to Purkinje fibres. Unfortunately, that limited our assessment to define the prevalence of non-Purkinje related ISCVT. The comparative analysis between CA and Pal/Syn groups were based on available ECGs to assess the outcome of the coinciding PVC on the T-wave. A few patients in the cohort had had cardiac MRI, RV biopsy and genetic testing.

6. Conclusion

Substantial inconsistency in literature exists over regarding the description of ventricular tachycarrhythmias in structurally normal hearts. Therefore, ISCVT as a new diagnostic term may help to unify these descriptions. ECG indices, in particular CI and CI/QT demonstrated good discriminatory performance to distinguish ISCVT from LCTDP.

Conflict of interest

We would also like to state that we have no conflicts of interest to declare.

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