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1 **Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) Mimics: The Knot**
2 **Unravelling By Cardiovascular Magnetic Resonance**

3

4 **Keywords:**

5 Arrhythmogenic Right Ventricular Cardiomyopathy; Magnetic Resonance Imaging; Heart; Cardiomyopathies;
6 Echocardiography

7

8 **Abbreviations:**

9 Arrhythmogenic right ventricular cardiomyopathy (ARVC); cardiovascular magnetic resonance (CMR); right
10 ventricle (RV); right ventricular end-diastolic volume (RVEDV); right ventricular end-systolic volume
11 (RVESV); right ventricular stroke volume (RVSV); right ventricular ejection fraction (RVEF); left ventricle
12 (LV); left ventricular ejection fraction (LVEF); left ventricular end-diastolic volume (LVEDV); left ventricular
13 end-systolic volume (LVESV); implantable cardioverter defibrillator (ICD); sudden cardiac death (SCD); Task
14 Force Criteria (TFC); late gadolinium enhancement (LGE); body surface area (BSA); transthoracic
15 echocardiogram (TTE); ischemic heart disease (IHD); atrial septal defect (ASD); arrhythmogenic left
16 ventricular cardiomyopathy (ALVC); left ventricular non compaction (LVNC).

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25 **Introduction:**

26 Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a rare genetic disease, with variable penetrance
27 [1]. First described in 1736 by Giovanni Maria Lancisi in “De Motu Cordis et Aneurysmatibus” [2], it was
28 initially thought to involve primarily the right ventricle (RV), with partial or total absence of the RV
29 musculature and fibro-fatty replacement [3-4], but recent evidence showed that in up to 70% of cases there is
30 also left ventricular (LV) involvement [5-7]. Clinical symptoms are often heterogeneous and non-specific,
31 including palpitations, syncope and atypical chest pain, hence representing a diagnostic challenge. ARVC can
32 lead to biventricular heart failure and sudden cardiac death (SCD), which represents the first manifestation of
33 the disease in up to 20% of cases [8]. Implantable Cardioverter Defibrillator (ICD) decreases the risk of SCD,
34 so a correct diagnosis is crucial. The diagnosis of ARVC is based on the 2010 Task Force Criteria (TFC) [9],
35 which recommend a multi-parametric approach that takes into account echocardiographic, electrocardiographic
36 and histologic abnormalities, documented ventricular arrhythmia and family history. Imaging criteria for ARVC
37 subtend potential diagnostic pitfalls of which the clinician needs to be aware: normal variants mischaracterized
38 as ARVC, such as chest wall deformity and non-ARVC fatty infiltration (obesity, post-myocardial infarction),
39 and pathologic conditions mimicking ARVC, such as myocarditis, sarcoidosis and pre-tricuspid shunts, which
40 are commonly referred to as ARVC mimics [10]. Cardiovascular Magnetic Resonance (CMR) as part of the
41 2010 TFC is increasingly used in clinical practice in patients with suspected ARVC in the context of a multi-
42 modality imaging assessment. The aim of our study was to assess the diagnostic role of CMR in patients
43 referred for suspected ARVC and its ability to identify ARVC mimics, and to explore its additional clinical
44 impact.

45

46 **Materials and methods:**

47 We retrospectively analysed the CMR registry data from the year 2014 (January to December) of a UK tertiary
48 centre, to identify consecutive patients referred for suspected ARVC. Clinical, ECG and echocardiographic data
49 were collected from clinical records. CMR was performed on a 1.5 T scanner (Magnetom Avanto, Siemens
50 Medical Solutions, Erlangen, Germany) and all patients underwent a CMR protocol including the left
51 ventricular (LV) and right ventricular (RV) anatomy, cine and late gadolinium enhancement (LGE) images.
52 Cine images were performed using a steady-state free-precession sequence in the 4-chamber, 3-chamber and 2-
53 chamber long-axis view, followed by a stack of short-axis slice from base to apex; typical image parameters

54 were TR 38 ms, TE 1.07 ms, flip angle 80°, bandwidth 930 Hz/Px, voxel size 2.0x2.0x8.0mm, slice thickness 8
55 mm, inter-slice gap 0 mm. Additional RVOT cine images were obtained, followed by a stack of axial views
56 (slice thickness 5mm, inter-slice gap 5mm) through the RVOT from the pulmonary valve to the RV
57 diaphragmatic wall. LGE images were obtained 15-20 minutes after intravenous administration of 0.1 mmol/Kg
58 of gadobutrol (Gadovist 1.0 mmol/ml, Bayer-Schering, Berlin, Germany) in identical planes to the long- and
59 short-axis cine images, using an inversion recovery segmented gradient echo sequence. Typical image
60 parameters were TR 700 ms, TE 3.15 ms, flip angle 25°, slice thickness 8.0 mm, interslice gap 0 mm, bandwidth
61 140 Hx/Px, voxel size 2.0x1.5x8.0 mm. The inversion time was progressively optimized to null normal
62 myocardium (typical values, 250–350 ms). Each slice was obtained during a breath-hold of 10–15 s depending
63 on the patient's heart rate. According to 2010 Task Force Criteria [9], CMR criteria were defined as *major* in the
64 presence of regional RV akinesia/dyskinesia/dyssynchronous contraction, associated with ratio of right
65 ventricular end-diastolic volume (RVEDV) to body surface area (BSA) ≥ 110 ml/m² (male) or ≥ 100 ml/m²
66 (female), or RV ejection fraction (RVEF) $\leq 40\%$; *minor* criterion was defined as the presence of regional RV
67 akinesia/dyskinesia/dyssynchronous contraction, associated with ratio of RV end-diastolic volume (RVEDV) to
68 body surface area (BSA) 100-109 ml/m² (male) or 90-99 ml/m² (female), or RV ejection fraction (RVEF) 40%-
69 45% . Body surface area was calculated using the Du Bois method. The study was reviewed by the local
70 Institutional Research and Innovation Department and in view of its retrospective design a formal ethical
71 approval was waived.

72

73 **Statistical analysis:**

74 Continuous and categorical variables were expressed as mean \pm SD and n (%), respectively. Continuous data
75 were compared by using the 2-tailed unpaired t test or by using the Mann-Whitney U test. Categorical variables
76 were compared by using the chi-square test or Fisher exact test, as appropriate. A p-value of <0.05 was
77 considered statistically significant; a Bonferroni-corrected p-value was used for comparison > 2 groups.
78 Comparisons between more than two groups were assessed using the Kruskal-Wallis test, using Dunn's test for
79 *post hoc* comparison. Data were analysed with SPSS® version 23 (IBM®).

80

81

82 **Results:**

83 Out of 2,481 scans performed in our CMR centre between Jan-Dec 2014, we identified 124 patients (5%) (56%
84 male, mean age 41 ± 16 years, age range 17-78 years) referred for suspected ARVC. Patients were referred with
85 suspected ARVC/D on the basis of symptoms, family history of ARVC and/or SCD, abnormal ECG or
86 abnormal transthoracic echocardiogram (TTE). Eighty-five patients (69%) were symptomatic: history of
87 palpitations/arrhythmias was reported in 53 patients (43%), syncope with no documented arrhythmia in 26
88 (21%) and both history of arrhythmia and syncope in 6 patients (5%), while thirty-nine patients (31%) were
89 asymptomatic, with an abnormal ECG and/or TTE found incidentally during school or competitive sport pre-
90 participation screening or pre-operatively. ECG data were available in 65 patients (52%): 53/65 patients (82%)
91 had abnormal ECG, most commonly T-wave inversion in leads V1-V3. Echocardiographic data were available
92 in 96 patients (77%): 26/96 patients (27%) had evidence of abnormal RV on echocardiogram. Family history of
93 SCD was reported in 16 patients (13%), 5 patients (4%) had family history of ARVC and one patient (1%) had
94 family history of both SCD and ARVC (**table 1**).

95

96 **CMR Findings:**

97 Biventricular volumes and function were overall preserved: mean LV ejection fraction (LVEF) was $61\pm 8\%$,
98 mean LV end-diastolic volume (LVEDV) was 83 ± 24 ml/m² and mean LV end-systolic volume (LVESV) was
99 34 ± 19 ml/m²; mean RV ejection fraction (RVEF) was $58\pm 8\%$, mean RVEDV 84 ± 23 ml/m² and mean RV end-
100 systolic volume (RVESV) was 36 ± 15 ml/m². Thirteen patients (10%) had evidence of LGE. Based on CMR
101 findings, a pathologic substrate was found in 36 patients (29%): ischemic heart disease (IHD) was found in 5
102 patients (4%) and non-ischemic heart disease in 10 (8%); 5 patients (4%) met CMR imaging criteria for ARVC
103 (**Figure 1A, B, C and D**), of which one had findings consistent with ALVC, and sixteen patients (13%) were
104 ARVC mimics. A structurally normal heart was found in 82 patients (66%) and non-specific findings (mild non-
105 specific regional wall motion abnormalities) in 6 (5%). CMR findings are listed in **Table 2**. Echocardiographic
106 data were available in 96 patients (77%). TTE and CMR findings agreed in 49 patients (51%); CMR provided
107 an entirely new diagnosis in 22 patients (22%) and found a structurally normal heart in 20 patients (21%) who
108 had abnormal findings on TTE. One patient (1%) was identified as ARVC mimics on TTE, as compared to 12
109 (13%) identified on CMR ($p=0.01$).

110 **ARVC mimics:**

111 Sixteen patients (13%) were found to have ARVC mimics on CMR. Six patients had normal variant
112 mischaracterized as ARVC: one patient had a pectus excavatum (**Figure 2A and B**) and five had findings
113 consistent with athlete's heart. Ten patients had pathologic conditions mimicking ARVC: cardiac sarcoidosis
114 (n=1), myocarditis (n=1), RV myocardial infarction (n=1), partial congenital absence of pericardium (n=1)
115 (**Figure 2C and D**); 3 patients were diagnosed with left ventricular non compaction (LVNC) and 3 with pre-
116 tricuspid left to right shunting (2 atrio-ventricular septal defect, ASD, and 1 partial anomalous venous return)
117 (**Figure 3**). There was no significant difference in clinical, ECG and TTE characteristics between patients with
118 structurally normal hearts on CMR and those with ARVC and ARVC mimics, and between ARVC and ARVC
119 mimics and the remaining population (Table 1). RVEDV and RV stroke volume (SV) were significantly higher
120 in patients with ARVC (RVEDV p=0.013, RVSV p=0.013) and ARVC mimics (RVEDV p=0.007, RVSV
121 p=0.012), as compared to those with structurally normal hearts. There was no significant difference in RV
122 volumes and function in patients with ARVC and ARVC mimics, while LVESV was significantly larger in
123 patients with ARVC. When comparing patients with ARVC and ARVC mimics (n=21) and the remaining
124 population (n=103), there was no significant difference in clinical, ECG and TTE characteristics while
125 biventricular volumes and RV stroke volume were significantly higher in patients with ARVC and ARVC
126 mimics (RVEDV 79 vs 103 ml/m², p=0.001; RVESV 34 vs 47 ml/m², p=0.018, RVSV 46 vs 56, p=0.001)(Tabel
127 2).

128 **Discussion:**

129 Arrhythmogenic right ventricular cardiomyopathy is a rare disease, with variable penetrance and prognosis.
130 Given the implications of such a diagnosis, the 2010 Task Force Criteria (TFC) recommended a multi-
131 parametric approach, comprehensive of imaging findings, family history, arrhythmias, ECG and histologic
132 abnormalities [9]. The symptoms of the disease are non-specific (chest pain, palpitations) and overlap with other
133 cardiomyopathies, thus not being helpful for a definite diagnosis [3,4,7,8]. It is well established that the
134 diagnosis of ARVC cannot rely on imaging findings alone, as imaging is subject to diagnostic pitfalls, such as
135 normal variants mischaracterized for ARVC (i.e. athlete's heart) or pathologic conditions mimicking it [10].
136 Bomma et al. [11] showed that less than 30% of patients referred for ARVC actually met the TFC after a
137 comprehensive clinical, invasive and non-invasive re-assessment. The advent of CMR offered a new insight into
138 ARVC [12-18]: due to its superior spatial resolution, unique tissue characterization, increased contrast between

139 blood pool and endomyocardium and multi-planarity, CMR is considered the gold standard for the assessment
140 of RV volumes and function. The implementation of the new TFC led to a significant reduction in the number of
141 patients confirmed with the diagnosis: Sen-Chowdhry reported an excellent sensitivity but low specificity (29%)
142 of CMR in relation to the TFC [19]. Similar findings were confirmed by Vermes et al. [20,21], which showed a
143 reduction in the prevalence of major and minor CMR criteria after the revised TFC. We found that only 5/124
144 patients (4%) referred for suspected ARVC actually met the TFC, in keeping with findings from Quarta et al.
145 [22] in a similar cohort. Normal and pathologic conditions mimicking ARVC make the diagnosis even more
146 challenging. Chest wall deformity and non-ARVC related fatty infiltration (obesity, lipomatous metaplasia post-
147 myocardial infarction) could be misinterpreted as ARVC. Moreover, increased RV volumes in athlete's heart or
148 pre-tricuspid shunting often lead to misdiagnosis [24-29]. In our study we observed that 16/124 patients (13%)
149 were found to have ARVC mimics, which were mainly represented by pathologic conditions rather than normal
150 variants mimicking the disease, leading to important clinical implications. In our cohort, the prevalence of
151 ARVC mimics was slightly higher compared with those previously reported in literature: Quarta et al. [22]
152 reported a 5% prevalence of ARVC mimics among patients referred to CMR for suspected ARVC, with similar
153 findings confirmed by Ting et al. [23], which showed a 4.4% prevalence of ARVC mimics. As CMR is part of
154 the multi-modality assessment in patients with suspected ARVC, it is increasingly used in clinical practice,
155 especially due to the potential clinical and prognostic implications that such a diagnosis would carry, and
156 sometimes it is performed to definitely rule out ARVC also in cases where pre-test likelihood is low; we think
157 this might at least in part explain the higher prevalence of ARVC mimics in our cohort. We also assessed the
158 ability of TTE to identify ARVC mimics, and found that CMR was significantly superior (13% by CMR vs 1%
159 by TTE, $p=0.01$). Although RV volumes were bigger in patients with ARVC and ARVC mimics, as compared
160 to the remaining population, the lack of difference among clinical, ECG, TTE and CMR characteristics between
161 ARVC and ARVC mimics, makes it challenging to identify ARVC mimics in the early differential diagnosis.
162 Interestingly, we also found that 82/124 patients (66%) with suspected ARVC based on clinical assessment
163 showed a structurally normal heart on CMR. Our study confirms and extends previous findings and highlights
164 the limitations of the TFC that do not consider the occurrence of ARVC mimics. Tissue characterization by
165 CMR, including LGE, might help in the differential diagnosis, however, to date, tissue characterization is
166 currently not included among the TFC. The main limitation of our study is the retrospective design; moreover
167 neither endomyocardial biopsy (given its little access in our Centre) nor genetic testing was available in our

168 cohort. As ARVC is a rare disease, prospective multicentre studies are needed to confirm and expand our
169 findings, aiming at improving the generalizability of our results.

170

171 **Conclusion:**

172 Out of 2,481 scans performed in our centre over a year, 124 (5%) were performed for suspected ARVC. Based
173 on CMR findings, a pathologic substrate was found in 29% of patients and a structurally normal heart in 66%.
174 ARVC imaging criteria were met in only 4% of patients, while 13% of patients showed findings consistent with
175 ARVC mimics. CMR showed to be superior to TTE in the identification of ARVC mimics (13% vs 1%, $p=0.01$)
176 and, overall, provided a change in diagnosis in 22% of patients. Accurate identification of the underlying
177 pathology in patients with suspected ARVC is pivotal given the impact on clinical management and prognosis.
178 Our study shows the incremental role of CMR in the identification of ARVC mimics, over and above TTE.

179

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184 of Health and Social Care.

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264

265

266

267 **Table 1.** Demographic and clinical characteristics.

268 **Table 2.** CMR findings.

269 **Figures captions**

270 **Figure 1. Right and left-dominant arrhythmogenic cardiomyopathy.**

271 Top panel. Diastolic (A) and systolic (B) four chamber view showing dilated right ventricle with bulging of the
272 free wall (solid arrows) in a patient meeting one major CMR criterion for ARVC. Bottom panel. Mid-cavity
273 short axis cine sequence (C) with evidence of right ventricular free wall late gadolinium enhancement (LGE) (D,
274 white arrow) and extensive LGE of the interventricular septum (C and D, black arrows) and left ventricular
275 inferolateral wall (D, white pentagon).

276

277 **Figure 2. Abnormal right ventricular features mimicking ARVC.**

278 Four chamber long axis cine view showing a distorted right ventricle (A) in a patient with pectus excavatum (B,
279 solid white arrow). Four chamber long axis cine view showing heart displacement towards the left with cardiac
280 apex pointing posteriorly (C, white arrow-head) and evidence of lung interposition between the aorta and the
281 pulmonary artery (D, white arrow) in a patient with partial congenital absence of the pericardium.

282

283 **Figure 3. Pre-tricuspid shunting mimicking ARVC**

284 Four chamber long axis view showing dilated right ventricle (A) in a patient with evidence of atrial septal defect
285 and left to right shunting on the short axis view (B, solid arrow). Four chamber long axis cine view showing
286 dilated right ventricle with septal flattening, in keeping with right ventricular overload (C) in a patient with left
287 upper pulmonary vein (D, white arrow) draining into the brachiocephalic trunk.