Case Report

Left Bundle Branch Block morphology Ventricular Tachycardia in a marathon runner

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A 41-year-old male presented to the accident and emergency department with a 3-hour history of palpitations and mild chest discomfort that had woken him from sleep. There was no history of previous episodes; indeed he had no past medical, surgical or family history of note.

He was an Iron Man marathon competitor.

The admission ECG showed a broad complex tachycardia (figure 1). Atrio-ventricular dissociation was evident with negative QRS concordance across the chest leads, fusion beats were also visible. The presence of a left bundle branch block, superior axis morphology further supported a diagnosis of ventricular tachycardia originating from the apex of the right ventricle.

Synchronized DC cardioversion was performed and his resting ECG was abnormal with extensive anteroinferior T wave inversion and Epilson waves in the leads III and aVF (figure 2).

On transthoracic echo the right ventricle was severely dilated and hypokinetic (figure 4). A coronary angiogram confirmed normal coronary arteries. On signal averaged ECG (SAECG), late low-amplitude ventricular potentials were evident.

The combined findings of sustained left bundle, superior axis ventricular tachycardia, repolarisation abnormalities on resting ECG, depolarisation abnormalities on Signal averaged ECG and regional structural abnormalities on 2d Echo fulfilled the revised Task Force criteria for diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) (Figure 3).

A cardiac MRI was performed and demonstrated the classical imaging findings of ARVC. The right ventricle was dilated, the apex and right ventricular (RV) outflow tract were dyskinetic and there was an aneurysmal segment in the basal inferior wall of the right ventricle. Abnormal gadolinium contrast enhancement was seen at the RV apex and also in the left ventricle reflecting disease severity and a higher risk of heart failure and arrhythmias (figure 5).

The patient was commenced on bisoprolol, and given the high future risk of sudden cardiac death, a dual chamber single coil defibrillator was implanted. Electrophysiological testing was not undertaken, as it has not been shown to be useful in risk stratification of ARVC.

Definitive diagnosis of ARVC is dependent on histological findings. However, transvenous endomyocardial biopsy has diagnostic limitations; the fibrofatty change of ARVC is often patchy and, as biopsies are taken usually taken from the septum rather than the RV free wall (to reduce the risk of perforation), diagnostic yield can be low. As the Taskforce diagnostic criteria had already been fulfilled in this case we elected not to undertake a biopsy.

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Fig 3. 2010 ESC revised Task Force Criteria for the Diagnosis of ARVC

| Major | By 2D echo: |
|-------|-------------|
| • Regional RV akinesia, dyskinesia, or aneurysm |
| • and 1 of the following (end diastole): |
| - PLAX RVOT ≥32 mm (corrected for body size [PLAX/BSA] ≥19 mm²/m²) |
| - PSAX RVOT ≥36 mm (corrected for body size [PSAX/BSA] ≥21 mm²/m²) |
| - or fractional area change <33 percent |

| Minor | By 2D echo: |
|-------|-------------|
| • Regional RV akinesia, dyskinesia, or aneurysm |
| • and 1 of the following (end diastole): |
| - PLAX RVOT ≥29 to <32 mm (corrected for body size [PLAX/BSA] ≥16 to <19 mm²/m²) |
| - PSAX RVOT ≥32 to <36 mm (corrected for body size [PSAX/BSA] ≥18 to <21 mm²/m²) |
| - or fractional area change ≥33 percent to ≤40 percent |

| Major | By MRI: |
|-------|---------|
| • Regional RV akinesia or dyskinesia or dyssynchronous RV contraction |
| • and 1 of the following: |
| - Ratio of RV end-diastolic volume to BSA ≥110 mL/m² (male) or ≥100 mL/m² (female) |
| - or RV ejection fraction <40 percent |

| Minor | By MRI: |
|-------|---------|
| • Regional RV akinesia or dyskinesia or dyssynchronous RV contraction |
| • and 1 of the following: |
| - Ratio of RV end-diastolic volume to BSA ≥100 to <110 mL/m² (male) or ≥90 to <100 mL/m² (female) |
| - or RV ejection fraction >40 percent to ≤45 percent |

| Major | By RV angiography: |
|-------|-------------------|
| • Regional RV akinesia, dyskinesia, or aneurysm |

| Minor | By RV angiography: |
|-------|--------------------|
| • Regional RV akinesia, dyskinesia, or aneurysm |

| 1. RV on Imaging |
|------------------|
| **Major** | **Minor** |
| By 2D echo: | By 2D echo: |
| • Regional RV akinesia, dyskinesia, or aneurysm | • Regional RV akinesia, dyskinesia, or aneurysm |
| • and 1 of the following (end diastole): | • and 1 of the following (end diastole): |
| - PLAX RVOT ≥32 mm (corrected for body size [PLAX/BSA] ≥19 mm²/m²) | - PLAX RVOT ≥29 to <32 mm (corrected for body size [PLAX/BSA] ≥16 to <19 mm²/m²) |
| - PSAX RVOT ≥36 mm (corrected for body size [PSAX/BSA] ≥21 mm²/m²) | - PSAX RVOT ≥32 to <36 mm (corrected for body size [PSAX/BSA] ≥18 to <21 mm²/m²) |
| - or fractional area change ≤33 percent | - or fractional area change ≥33 percent to ≤40 percent |

| 2. Histology |
|--------------|
| **Major** | **Minor** |
| Residual myocytes <60 percent by morphometric analysis (or <50 percent if estimated), with fibrous replacement of the RV free wall myocardium in ≥1 sample, with or without fatty replacement of tissue on endomyocardial biopsy | Residual myocytes 60 percent to 75 percent by morphometric analysis (or 50 percent to 65 percent if estimated), with fibrous replacement of the RV free wall myocardium in ≥1 sample, with or without fatty replacement of tissue on endomyocardial biopsy |

| 3. ECG - Repolarization abnormalities |
|-------------------------------------|
| **Major** | **Minor** |
| Inverted T waves in right precordial leads (V₁, V₂, and V₃) or beyond in individuals >14 years of age (in the absence of complete right bundle-branch block QRS ≥20 ms) | Inverted T waves in leads V₁, V₂, V₃, and V₄ in individuals >14 years of age in the presence of complete right bundle-branch block |

| 4. ECG - Depolarization/conduction abnormalities |
|-----------------------------------------------|
| **Major** | **Minor** |
| Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right precordial leads (V₁ to V₄) | Late potentials by SAECG in ≥1 of the following 3 parameters in the absence of a QRS duration of ≥110 ms on the standard ECG: |
| • Filtered QRS duration (fQRS) ≥114 ms |
| • Duration of terminal QRS <40 µV (low-amplitude signal duration) ≥38 ms |
| • Root-mean-square voltage of terminal 40 ms ≤20 µV |
| • Terminal activation duration of QRS ≥55 ms measured from the nadir of the S wave to the end of the QRS, including R⁺, in V₁, V₂, V₃, or V₄, in the absence of complete right bundle-branch block |

| 5. Arrhythmias |
|----------------|
| **Major** | **Minor** |
| Nonsustained or sustained ventricular tachycardia of left bundle-branch morphology with superior axis (negative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL) | Nonsustained or sustained ventricular tachycardia of left bundle-branch morphology with superior axis (negative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL) |

| 6. Family History |
|--------------------|
| **Major** | **Minor** |
| ARVC/D confirmed in a first-degree relative who meets current Task Force criteria | History of ARVC/D in a first-degree relative in whom it is not possible or practical to determine whether the family member meets current Task Force criteria |
| ARVC/D confirmed pathologically at autopsy or surgery in a first-degree relative | Premature sudden death (<35 years of age) due to suspected ARVC/D in a first-degree relative |
| Identification of a pathogenic mutationΔ categorized as associated or probably associated with ARVC/D in the patient under evaluation | ARVC/D confirmed pathologically or by current Task Force Criteria in second-degree relative |

Definite diagnosis = 2 Major or 1 Major and 2 Minor criteria or 4 Minor from different categories

Borderline Diagnosis = 1 Major and 1 Minor or 3 Minor criteria from different categories

Possible Diagnosis = 1 Major or 2 Minor criteria from different categories

PLAX: parasternal long-axis view; RVOT: RV outflow tract; BSA: body surface area; PSAX: parasternal short-axis view; aVF: augmented voltage unipolar left foot lead; aVL: augmented voltage unipolar left arm lead.
He has been referred to medical genetics and is undergoing genetic testing and family screening.

The diagnosis had major implications for his life as he had been advised to cease all high intensity activity.

**DISCUSSION**

ARVC is an inherited cardiomyopathy characterised by RV dilation, thinning & dysfunction. Histologically, the RV myocardium is replaced by fibrous fatty tissue. It is a disease of the desmosomes; the mechanical connections between myocytes are disrupted hence the predilection for thin RV and the observation that the disease is more severe and presents earlier in athletes.

The prevalence of ARVC is estimated at 1:1000. It is an autosomal dominant condition with variable penetrance.

Most patients present between the ages of 10-50 years. The main presenting symptoms of ARVC are dizziness, palpitations, atypical chest pain and syncope. It is estimated that ARVC accounts for 20% of sudden cardiac deaths with an overall mortality rate of 2.5% per year.

A definite diagnosis of ARVC requires the presence of a combination of major and minor clinical criteria as set out by the 2010 ESC revised Task Force Criteria.

Many patients are asymptomatic and the diagnosis is often only considered due to non-specific ECG or echocardiographic abnormalities of the right ventricle. Clinicians are encouraged to consider a diagnosis of ARVC in these patients and to refer on to cardiology for further investigation.

VT with a left bundle branch block morphology and an inferior axis commonly originates from the RV outflow tract (RVOT). In contrast to Arrhythogenic Right Ventricular Cardiomyopathy, RVOT VT occurs in structurally normal hearts (occasionally the RVOT is dilated and RV regional wall motion abnormalities are seen on CMR) and is readily treatable with verapamil and betablockers or radiofrequency ablation. The ECG in sinus rhythm in RVOT VT is normal as is the SAECG. In contrast to ARVC, there are no family screening implications with RVOT VT.

Other differentials to consider include idiopathic dilated cardiomyopathy (IDCM) and Uhl’s anomaly. Patients with IDCM usually have a progressive decline in left ventricular function, in contrast to ARVC where the right heart is primarily affected. In Uhl’s anomaly the RV myocardium is paper thin and devoid of myocardium. There is no replacement of muscle by fatty tissue. It usually presents in childhood.

**CONCLUSION**

ARVC is a recognised cause of sudden cardiac death. Left bundle branch block morphology ventricular tachycardia or an abnormal right ventricle on echo in an otherwise well patient should prompt onward referral to cardiology.
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