Arrhythmia from RV; Benign vs Malignant?

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RV Arrhythmia

Primary event vs. Secondary phenomenon

BENIGN

Structural heart disease
Cardiomyopathy

Electrical disease
Malignant PVCs, which we define as the ability of idiopathic ventricular extrasystoles to either trigger lethal arrhythmias or induce cardiomyopathy.
Characteristic of a malignant, arrhythmogenic PVC is that the initiating beat that triggers PMVT/VF has a similar morphology to isolated ectopic beats that precede or follow the index event.
PVC

**PRIMARY**
- PVC induced cardiomyopathy
- Trigger of ventricular fibrillation

**SECONDARY**
- Electrical disease
- Structural heart disease
  - Cardiomyopathy
PVC induced Cardiomyopathy

Mechanism

1. Promotion of ventricular dyssynchrony
2. Increased oxygen consumption
3. Autonomic dysregulation
4. Impaired contractility with altered intracellular calcium currents
Risk Factors for PVC induced CMP

① high PVC frequency, (particularly >20%)
② epicardial PVC location,
③ Non-outflow tract origin
④ duration of PVC exposure, a gradual progression of PVC-CMP over 4–8 years.
⑤ QRS duration; QRS duration ≥150 ms.
⑥ Interpolated PVCs, nonsustained VT burden, short PVC coupling interval
⑦ male gender,
⑧ absence of circadian fluctuation of the PVC burden,
⑨ asymptomatic status.
Dyssynchrony

RV vs LV
RV Free wall vs RV Septum

Primary sources of malignant PVCs

The two primary sources

1. the Purkinje system and its distal arborized fibers
2. the myocardium of the RV and LV outflow tracts
Morphology of malignant PVCs

**RV**
- The morphology of PVC from RV Purkinje origin from moderator band
  - ① left bundle branch block (LBBB),
  - ② left axis deviation
  - ③ late precordial transition (>V4)
- The coupling interval of the PVC that triggered VF was usually (but not always) <300 ms.

**LV**
- Malignant PVCs originating from the LV Purkinje system localize along the ventricular septum and morphologically resemble fascicular beats, presenting with
  - ① a relatively narrow QRS complex,
  - ② a RBBB configuration
  - ③ a superior, inferior, or intermediate axis.

Sources of malignant PVCs
Illustrative Case of malignant RVOT PVC.
① If a developing cardiomyopathy is in a subclinical, occult phase during its transition to a manifest form, standard imaging modalities such as echocardiography may not detect any abnormalities.

② Cardiac MRI may be particularly helpful in distinguishing between an early phase of arrhythmogenic right ventricular cardiomyopathy (ARVC) and idiopathic RVOT since a majority of patients with idiopathic RVOT ectopy have structurally normal hearts.

③ Cardiac MRI and PET should be considered in those suspected of having cardiac sarcoidosis.
Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)

ARVC prevalence ranging between 1 in 2500 and 5000.

ARVC is an inherited cardiomyopathy with an autosomal dominant pattern of inheritance, incomplete penetrance, and variable expressivity.

A pathogenic mutation can be identified in approximately two-thirds of those diagnosed with the disease.

The mutations most commonly involve the desmosomal proteins.

The pathological hallmark of ARVC is myocyte loss with fibrofatty replacement of the myocardium.

Formation of scar begins in epicardial portions of the RV and progresses to the endocardium.
**ARVC DIAGNOSIS**

### Aberrant repolarization
- TWI in V₁ & V₂ or V₄, V₅ or V₆ in pts. >14 years old
- TWI in V₁, V₂, V₃ & V₄ in pts. >14 years old with complete RBBB Signal-average Electrocardiogram

### Aberrant depolarization
- Filtered QRS ≥114 ms
- OR 1 of the following
  - Terminal duration of QRS ≥55 ms
  - Terminal duration of QRS ≥38 ms (if <40 μV)
  - Root-mean-square voltage of terminal 40 ms of QRS <20 μV
  - NSVT or sustained VT LBBB morphology and inferior axis
  - >500 PVCs in 24-h period

### Arrhythmia history
- NSVT or sustained VT with LBBB morphology and superior axis
- 1st degree relative with confirmed ARVC by Task force criteria
- 1st degree relative with pathologic confirmation of ARVC on autopsy
- Known pathogenic mutation

### Arrhythmia Family history
- NSVT or sustained VT with LBBB morphology and superior axis
- 1st degree relative with confirmed ARVC by Task force criteria
- 1st degree relative with confirmed ARVC by Task force criteria
- 1st degree relative <35 years old with SCD
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<th>Emerging Diagnostic tools for ACM. Derived from Gandjbakhch et al. [1].</th>
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<td><strong>ECG</strong></td>
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<td>QRS fragmentation</td>
<td>Hypertrophic trabecular or hyper-reflective moderator band</td>
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<td>V1V2V3 QRS duration ≥110 &lt;ms</td>
<td>Decreased TAPSE and peak systolic RV annular velocity</td>
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<td>V1 + V2 + V3/V4 + V5 + V6 widths ≥1.2</td>
<td>Intra-myocardial fat infiltration in the RV wall</td>
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<td>V1V2V3 QRS width ≥25 ms of V6 (parietal block)</td>
<td>LGE of the RV wall</td>
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<td>S-wave upstroke ≥55 ms</td>
<td>Low peak systolic RV strain</td>
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<td>QRS fragmentation</td>
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<td>Inverted T waves in inferior leads (LV extension)</td>
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<td>RBBB with R'/S ratio &lt;1 in V1</td>
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Abbreviations: TTE, transthoracic echocardiograph; RV, right ventricular; MRI, magnetic resonance imaging; MDCT, multi-dimensional computed tomography; CT, computed tomography; RBBB, right bundle branch block; LV, left ventricular; LGE, late gadolinium enhancement; VA, ventricular arrhythmia; TAPSE, tricuspid annular plane systolic excursion; PVC, premature ventricular contraction; NSVT, non-sustained ventricular tachycardia.
Impact of Sports Activity on ACM
Exercise has been shown to have greater effect on the RV than the LV, causing wall stress on the RV many times greater than that of the LV, resulting in RV remodeling over time.

RV dilation, RV dysfunction, and elevation in cardiac biomarkers have been shown to occur acutely in endurance athletes after prolonged exercise.

These acute and chronic, stress-induced changes to the RV myocardium may result in the formation of an arrhythmogenic substrate.

There are two ways to interpret these observations.

① The first is that EC is a distinct entity from conventional ARVC.

② The second perspective is that these patients represent a mutation-negative and family history-negative subset of ARVC.
RV PVCs are malignant if they have an ability of idiopathic ventricular extrasystoles

① to either trigger lethal arrhythmias or induce cardiomyopathy

② secondary phenomenon of underlying disease (structural or electrical)

Recognition of these entities is critical for they may erroneously be attributed to an underlying cardiomyopathy with an irreversible pathogenesis.
We are constantly misled by the ease with which our minds fall into the ruts of one or two experiences.

-Sir William Osler

Thank you