Arrhythmogenic Cardiomyopathy versus Idiopathic Right Ventricular Outflow Tract Tachycardia

Denominations:

ACM: arrhythmogenic cardiomyopathy (the newest designation); **ARVC**: arrhythmogenic right ventricular cardiomyopathy; **ARVD**: arrhythmogenic right ventricular dysplasia; **ARVC/D**: arrhythmogenic right ventricular cardiomyopathy/dysplasia; **LDAC**: Left-dominant arrhythmogenic cardiomyopathy; and **ALVC**: Arrhythmogenic left ventricular cardiomyopathy.

ACM versus idiopathic Right ventricular outflow tract tachycardia (RVOT)

Differentiation between early-phase ACM and RVOT-VT can be challenging, and correct diagnosis is relevant. RVOT-VT is a form of monomorphic VT originating from the RVOT or occasionally from the tricuspid annulus. It is usually seen in patients without SHD. The two predominant causes of RVOT-VT are RVOT or idiopathic VT arising from the RVOT and ACM. The differential diagnosis between these 2 entities is critical, as their prognoses and therapeutic options differ. An incorrect diagnosis may be devastating. Both of these arrhythmias can be adrenergically mediated and may be difficult to distinguish clinically. RVOT is a clinical arrhythmic condition that is not typically associated with SHD as is seen in ACM. ECG and CMRI may be useful to distinguish these disorders.

The microvolt T-wave alternans (TWA or MTWA) is widely used to predict lethal ventricular arrhythmias in various diseases. However, the clinical significance of TWA in patients with VT originating from the RV has been unknown. TWA refers to beat-to-beat fluctuations of T-wave amplitude and morphology, and is associated clinically with impending ventricular arrhythmias and increased risk of SCD. TWA analysis can be done as part of an exercise stress test or during a Holter monitoring recording. Within candidates for ICD therapy, a negative TWA test may be useful in identifying low-risk patients who are unlikely to benefit from ICD placement. However, currently there is not enough evidence to support the use of TWA in clinical practice to guide therapy(**Aro AL, Kentta TV, Huikuri HV. Microvolt T-wave Alternans: Where Are We Now? Arrhythm Electrophysiol Rev. 2016; 5:37-40. doi: 10.15420/aer.2015.28.1**).

Yalin et al aim to investigate the possible role of TWA to discriminate ACM from idiopathic RVOT-VT. They enrolled 38 patients (23 males, 43 ± 16 years) with VT originating from the RV. TWA was measured during exercise testing using a modified moving average method. TWA results were compared among patients with ACM and RVOT-VT. Twenty-five patients (16 males, 42 ± 16 years) met the Task Force 2010 criteria for the diagnosis of ACM, and 13 patients (7 males, 45 ± 14 years) had idiopathic RVOT-VT. Twenty patients with ACM had positive TWA test, whereas only 1 patient with RVOT-VT had (80% versus 8%, P < 0.001). In patients with VT of RV origin, positive TWA test supports the diagnosis of ACM.(**Yalin K, Golcuk E, Aksu T, Tiryakioglu SK, Bilge AK, Adalet K. Distinguishing Right Ventricular Cardiomyopathy From Idiopathic Right Ventricular Outflow Tract Tachycardia with T-wave Alternans. Am J Med Sci. 2015; 350:463-6. doi: 10.1097/MAJ.0000000000000590**) Absence of significant TWA in a patient with cardiac disease with CHF, low ejection fraction or a recent MI is associated with a low risk of SCD. The following sequence shows the main differences between ACM with Idiopathic RVOT-VT

Differential diagnosis between ACM and Idiopathic RVOT-VT

I) Prevalence

- **ACM:** The estimated prevalence of ACM in the general population ranges from 1 in 1,000 to 1 in 5,000.(**Gemayel C, Pelliccia A, Thompson PD. Arrhythmogenic right ventricular cardiomyopathy. J Am Coll Cardiol. 2001; 38:1773-81. doi: 10.1016/s0735-1097(01)01654-0**) Peters refers 1: 1,000 to 1: 1,250 or 1:2000–1:5000.
- **Idiopathic RVOT-VT:** There was a high prevalence of J-waves in the idiopathic RVOT-VT/PVC patients referred for RFCA. Although patients with idiopathic RVOT arrhythmias associated with J-waves might have a more enhanced arrhythmogenicity than those without J-waves, the significance of those J-waves was limited in terms of the prognosis and VF. (**Yoshihiro Yamashina et al. Prevalence and characteristics of idiopathic right ventricular outflow tract arrhythmias associated with J-waves. Europace. 2011 Dec;13(12):1774-80. doi: 10.1093/europace/eur256.)**

II) Presence of gene mutation and inheritance pattern

- **ACM:** gene mutation ≈ 80% positive predominantly in desmosomes with inheritance pattern autosomal dominant (AD), autosomal recessive (AR), *Compound Heterozygosity or Digenic Mutations.* Gene mutations have been found in about 60% of people with ACM. Mutations in a desmosomal gene PKP2(**plakophilin-2**) appear to be most common. Of patients with plakophilin-2 genetic variants, 25 of 38 (65.7%) were found to have a second plakophilin-2 abnormality or a second abnormal desmosomal gene. (**Frank I Marcus et al. Genetics of arrhythmogenic right ventricular cardiomyopathy: a practical guide for physicians. J Am Coll Cardiol. 2013 May 14;61(19):1945-8. doi: 10.1016/j.jacc.2013.01.073**. In people without an identified mutation, the cause of the disorder is unknown. Researchers are looking for additional genetic factors that play a role in causing ACM. ACM confirmed in a first-degree relative who meets current TFC; ACM confirmed pathologically at autopsy or surgery in a first-degree relative Identification of a pathogenic mutation categorized as associated or probably associated with ACM in the patient
- **Idiopathic RVOT-VT:** No. They are the most common subtype of idiopathic ventricular arrhythmias. Research conducted under the leadership of Weill Cornell Cardiology Professor Bruce Lerman since the 1980s has suggested that RVOT is caused by a mutation in the gene for a protein called Gsα, and indeed such a mutation has been discovered in cardiac cells in Dr. Lerman's laboratory. (**Bruce B Lerman. Outflow tract ventricular arrhythmias: An update Trends Cardiovasc Med. 2015 Aug;25(6):550-8. doi: 10.1016/j.tcm.2015.01.011**.)

III) Sex M/F ratio

- **ACM:** M/F 70%/30%. M/F 3:1 (**Nava *A, et al.* Clinical profile and long-term follow-up of 37 families with arrhythmogenic right ventricular cardiomyopathy. *J Am Coll Cardiol. 2000 Dec; 36(7):2226-33***.). Sex-based differences in the incidence are conflicting. Italian studies report that it is more common in males with an approximate ratio of 3:1, (**Bauce B, Frigo G, Marcus FI, *et al.* Comparison of clinical features of arrhythmogenic right ventricular cardiomyopathy in men versus**

women. Am J Cardiol. 2008;102:1252-7. doi: 10.1016/j.amjcard.2008.06.054) on the other hand the United States and the Dutch ACM cohorts report similar incidence between males and females.(**Groeneweg JA, Bhonsale A, James CA, *et al.* Clinical Presentation, Long-Term Follow-Up, and Outcomes of 1001 Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy Patients and Family Members. Circ Cardiovasc Genet. 2015;8:437-46. doi: 10.1161/CIRCGENETICS.114.001003**) (**Bhonsale A, Groeneweg JA, James CA, *et al.* Impact of genotype on clinical course in arrhythmogenic right ventricular dysplasia/cardiomyopathy-associated mutation carriers. Eur Heart J. 2015;36:847-55. doi: 10.1093/eurheartj/ehu509**)European studies (predominantly from the Netherlands) and the United States show that AMC is 1.2–3 times more common in males (**Corrado et al., et al. . (1997). Spectrum of clinic pathologic manifestations of arrhythmogenic right ventricular cardiomyopathy/dysplasia: a multicenter study. *J. Am. Coll. Cardiol.* 30, 1512–1520. 10.1016/S0735-1097(97)00332-X**)(**Bauce B., et al. (2008). Comparison of clinical features of arrhythmogenic right ventricular cardiomyopathy in men versus women. *Am. J. Cardiol.* 102, 1252–1257. 10.1016/j.amjcard.2008.06.054**) (***Cox MG, et al..,*. **(2011). Arrhythmogenic right ventricular dysplasia/cardiomyopathy: pathogenic desmosome mutations in index-patients predict outcome of family screening: Dutch arrhythmogenic right ventricular dysplasia/cardiomyopathy genotype-phenotype follow-up study. *Circulation* 123, 2690–2700. 10.1161/CIRCULATIONAHA.110.988287**) (**Bhonsale et al., 2015 Impact of genotype on clinical course in arrhythmogenic right ventricular dysplasia/cardiomyopathy-associated mutation carriers. *Eur. Heart J.* 36, 847–855. 10.1093/eurheartj/ehu509)** ; (**Groeneweg et al., 2015 (2015). Clinical presentation, long-term follow-up, and outcomes of 1001 arrhythmogenic right ventricular dysplasia/cardiomyopathy patients and family members. *Circ. Cardiovasc. Genet.* 8, 437–446. 10.1161/CIRCGENETICS.114.001003**). However, the male

predominance is not observed in USA Registry (89% of males vs. 84% of females)

- **Idiopathic RVOT-VT:** M/F 24%/76% male/female ratio 0.49(**Mikiko Nakagawa et al, Gender differences in various types of idiopathic ventricular tachycardia J Cardiovasc Electrophysiol. 2002 Jul;13(7):633-8. doi: 10.1046/j.1540-8167.2002.00633.x**.)

IV) Age of presentation

- **ACM:** Early-phase ACM patients are younger than the RVOT-VT patients, a result of the early detection of ACM mutation positive by family screening.
- **Idiopathic RVOT-VT:** Typically seen between 20 and 50 years of age.

V) Geographic distribution

- **ACM:** Worldwide (Italy/Padua. Naxos island AR variant. The non desmosomal gene TMEM43-endemic to Newfoundland, Canada. To date, all familial cases reported worldwide share a common ancestral haplotype. (**Dominguez F, Zorio E, Jimenez-Jaimez J, *et al.* Clinical characteristics and determinants of the phenotype in TMEM43 arrhythmogenic right ventricular cardiomyopathy type 5. Heart Rhythm. 2020;17:945-54. doi: 10.1016/j.hrthm.2020.01.035**)
- **Idiopathic RVOT-VT:** Worldwide.

VI) Main clinical manifestations

- **ACM:** Asymptomatic (6.2%), Palpitations (67%), Exertional pre-syncope, syncope (32%), atypical chest pain (27%) or SCD. Syncope: more prevalent. Not unexpectedly, SCD/CA: more prevalent.
- **Idiopathic RVOT-VT:** Asymptomatic (26,6%). palpitation (30%), presyncope (43,4%) lightheadedness often provoked by sympathetic stimulation during exercise or emotional upset. Syncope: occasionally observed. Lesser prevalent. Not unexpectedly, SCD/CA: Rare.

VII) ECG manifestations

- **ACM: Depolarization features:** QRS duration in right precordial leads ≥105 ms registered in≈ 80% of case; TAD of QRS ≥55 ms: Present in ≈30% of cases, specificity of 100%. Excellent accuracy.; *Epsilon wave:*

Present in 30% of cases with conventional 12-leads ECG, in the right precordial leads (Mayor criteria?). Its identification and interpretation are influenced by ECG filtering and sampling rate, with large interobserver variability (**Platonov PG, et al. High interobserver variability in the assessment of epsilon waves: Implications for diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia. Heart Rhythm . 2016 Jan;13(1):208-16. doi: 10.1016/j.hrthm.2015.08.031**.). Consequently, currently Padua researches, consider epsilon waves in right precordial leads ***a minor ECG criterion***.

- **Repolarization features:** JT interval duration in right versus left precordial leads ≥1.15: Yes. High specificity = 97%.; TWI from V1 to V3 during sinus rhythm in absence of IRBBB or CRBBB is registered in 15% of cases: specificity very high and moderate sensitivity. TWI typically remain inverted during exercise (**Bauce B, Frigo GMP, Benini G, et al. Differences and similarities between Arrhythmogenic right ventricular cardiomyopathy and athlete's heart adaptions. *Br J Sports Med.* 2008**)
- **Idiopathic RVOT-VT: Depolarization features:** QRS duration in right precordial leads ≥105 ms absent or rare. TAD of QRS ≥55 ms: Absent.; Epsilon wave**:** Absent.; **Repolarization features:** JT interval duration in right versus left precordial leads ≥1.15: Absent.; TWI from V1 to V3 during sinus rhythm: rare: 1% of cases. TWI typically normalize during exercise among athletes (***Nasir K, 2004*** **Electrocardiographic features of arrhythmogenic right ventricular dysplasia/cardiomyopathy according to disease severity: a need to broaden diagnostic criteria *Circulation. 2004 Sep 21;110(12):1527-34. doi: 10.1161/01.CIR.0000142293.60725.18.***).

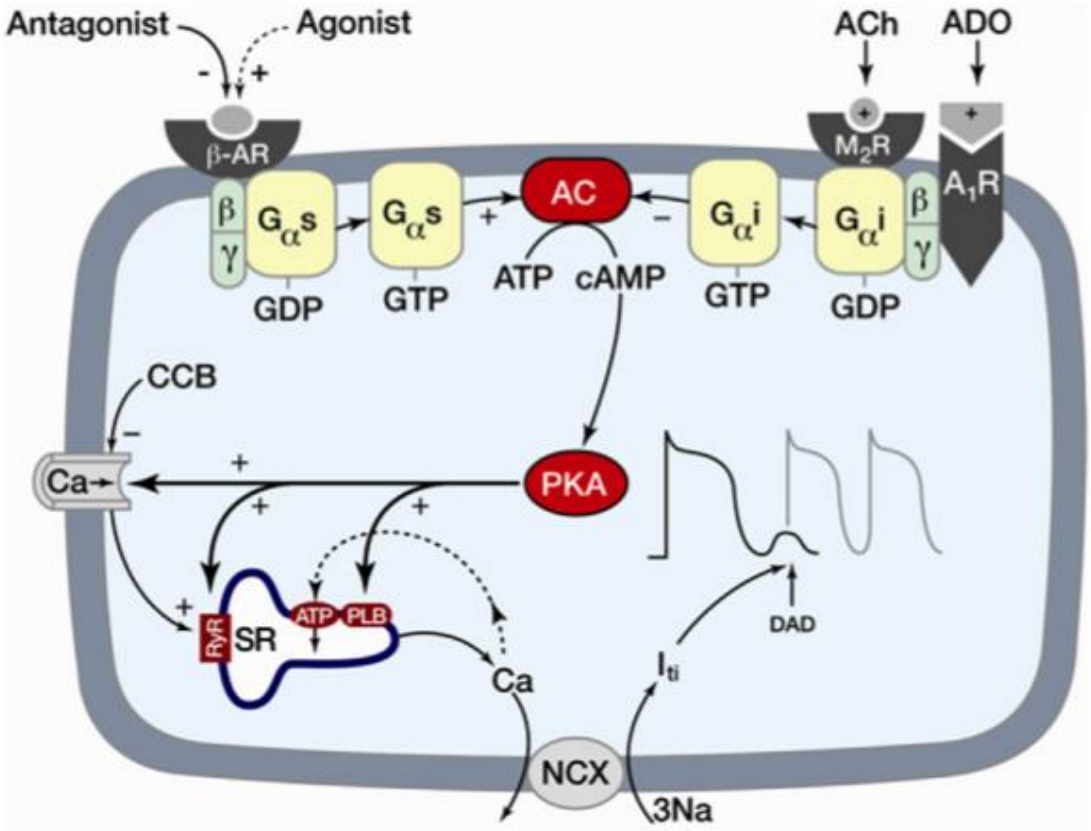
VIII) Ventricular arrhythmias

- **ACM:** PVCs frequency: Relatively low number: Less frequent PVCs and NSVT in early-phase.; PVCs v of RVOT: In early-phase ACM frequent origin of PVC mainly the **septal** part of the RVOT (98%). (**Jørg Saberniak, et al. Comparison of patients with early-phase arrhythmogenic right ventricular cardiomyopathy and right**

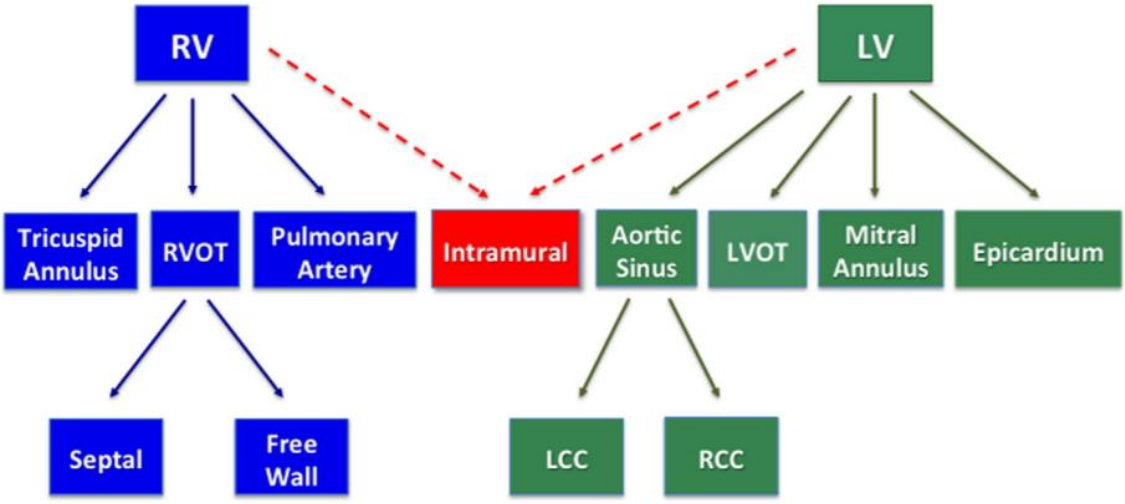
ventricular outflow tract ventricular tachycardia. Eur Heart J Cardiovasc Imaging. 2017 Jan; 18(1): 62–69. doi: 10.1093/ehjci/jew014).; Polymorphic VT and VF: More common and generally familial.; **Morphology of monomorphic VT**: NSVT or SVT of LBBB morphology with superior axis (negative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL). It is considered a Mayor criterion.; Mechanism of VT: Reentry originate from fibro-fatty replacement of myocardium forming the substrate for VT. In early phase, other mechanisms may be involved including disruptive electrical conduction in addition to early fibrosis. (***Corrado D, et al.* Molecular biology and clinical management of arrhythmogenic right ventricular cardiomyopathy/dysplasia. *Heart. 2011 Apr; 97(7):530-9*.**). Event triggers: During exercise. ACM accounts for up to 20% of cases of SCD in young athletes (**Corrado D., et al (1998). Screening for hypertrophic cardiomyopathy in young athletes. *N. Engl. J. Med.* 339, 364–369. 10.1056/NEJM199808063390602**) (**Tabib A., et al. (2003). Circumstances of death and gross and microscopic observations in a series of 200 cases of sudden death associated with arrhythmogenic right ventricular cardiomyopathy and/or dysplasia. *Circulation* 108, 3000–3005. 10.1161/01.CIR.0000108396.65446.21**)

- **Idiopathic RVOT-VT:** PVCs frequency: More frequent. PVCs predominant, site of origin of PVC in the **lateral** free wall of RVOT (***Zhang F et al.* Electrocardiographic algorithm to identify the optimal target ablation site for idiopathic right ventricular outflow tract ventricular premature contraction., *Europace. 2009 Sep; 11(9):1214-20*.**).; Polymorphic VT and VF: Rare. VTs are monomorphic and generally not familial. **Morphology of monomorphic VT**: NSVT of LBBB morphology with inferior axis (right or left) with tall R waves in leads II, III, and aVF. The arrhythmia may present occasionally with SVT, NSVT or PVCs, often provoked by exercise or emotional upset.; **Mechanism:** adenosine-sensitive, cyclic AMP mediated, triggered activity (**Lerman *BB* Mechanism, diagnosis, and treatment of outflow tract tachycardia. *Nat Rev Cardiol. 2015 Oct; 12(10):597-608*.**) (***Kim RJ, et al.* Clinical and**

electrophysiological spectrum of idiopathic ventricular outflow tract arrhythmias ***J Am Coll Cardiol. 2007 May 22; 49(20):2035-43***.). Figures



Mechanism of Outflow Tract Tachycardia. Signal transduction schema for initiation and termination of cAMP mediated triggered activity. AC ¼ adenylyl cyclase; ACh ¼ acetylcholine; ADO ¼ adenosine; A1R ¼ A1-adenosine receptor; β-AR ¼ β-adrenergic receptor; CCB ¼ calcium channel blocker; DAD ¼ delayed afterdepolarization; Iti—transient inward current; M2R ¼ muscarinic receptor; NCX ¼ Naþ/Ca2þ exchanger; PLB ¼ phospholamban; PKA ¼ protein kinase A; RyR ¼ Ryanodine receptor; SR ¼ sarcoplasmic reticulum. (Reproduced from Lerman.)



Classification of outflow tract tachycardia. LCC ¼ left coronary cusp; LV ¼ left ventricle; LVOT ¼ left ventricular outflow tract; RCC ¼ right coronary cusp; RV ¼ right ventricle; RVOT ¼ right ventricular outflow tract.

IX) Transthoracic Echocardiogram (TTE)

- **ACM:** Increased RV diameters, Additionally, RV function is decreased, and RV mechanical dispersion is pronounced. LV function is reduced. LVEF and Left ventricular global longitudinal strain (LVGLS), and LV mechanical dispersion is more pronounced.
- **Idiopathic RVOT-VT:** All RV diameters are within normal range in the RVOT-VT patients. RV function is in the lower normal range.

X) 3-dimensional Electro Anatomical Voltage Mapping (EVM) Right ventricular EVM identify low-voltage regions ("electroanatomical scar"), which in patients with ACM correspond to areas of fibrofatty myocardial replacement.

- **ACM:** An early/minor form of ACM may mimic idiopathic RVOT tachycardia. EVM is able to identify Idiopathic RVOT-VT due to concealed ACM by detecting RVOT electroanatomical scars that correlate with fibrofatty myocardial replacement at EMB and predispose to SCD. (**Corrado D et al, Three-dimensional electroanatomical voltage mapping and histologic evaluation of myocardial substrate in right ventricular outflow tract tachycardia J Am Coll Cardiol. 2008 Feb 19;51(7):731-9. doi: 10.1016/j.jacc.2007.11.027.)**

- **Idiopathic RVOT-VT:** Electroanatomical voltage mapping was normal in 74%, with electrogram voltage >1.5 mV throughout the RV. (**Corrado D et al, Three-dimensional electroanatomical voltage mapping and histologic evaluation of myocardial substrate in right ventricular outflow tract tachycardia J Am Coll Cardiol. 2008 Feb 19;51(7):731-9. doi: 10.1016/j.jacc.2007.11.027.).**

XI) Cardiac Magnetic Resonance Image (CMRI)

- **ACM:** Visualization of fibro-fatty infiltration on T1-weighted images.
- **Idiopathic RVOT-VT:** Is normal presence of fat in the AV groove and anteroapical RV epicardium. Artifacts due to motion, arrhythmia, and surface coil proximity can reduce specificity (**Asimaki A et al. A new diagnostic test for arrhythmogenic right ventricular cardiomyopathy. N Engl J Med. 2009 Mar 12;360(11):1075-84. doi: 10.1056/NEJMoa0808138**.) It is particularly important to exclude mild forms of arrhythmogenic right **ventricular dysplasia/cardiomyopathy**. Patients with a family history of SCD and apparent RVOT tachycardia should be particularly thoroughly with a CMRI.

XII) RV ejection fraction:

- **ACM:** Limited value
- **Idiopathic RVOT-VT:** RV Highly effective

XIII) Endomyocardial biopsy(EMB)

- **ACM:** Residual myocytes <60% by morphometric analysis (or <50% if estimated), with fibrous replacement of the RV free wall myocardium in ≥1 sample, with or without fatty replacement of tissue on endomyocardial biopsy
- **Idiopathic RVOT-VT:** Idiopathic ventricular tachycardia is defined as VT that occurs in patients without structural heart disease (SHD), metabolic abnormalities, or the LQTS.

XIV) Prognosis

- **ACM:** Tendency to ventricular arrhythmias, biventricular dysfunction, cardiac syncope, SCD and therefore a far from a benign condition. (Corrado ***D, et al.*** **Treatment of Arrhythmogenic Right Ventricular**

Cardiomyopathy/Dysplasia: An International Task Force Consensus Statement. *Circulation. 2015 Aug 4; 132(5):441-53*.)

- **Idiopathic RVOT-VT:** Usually benign, but occasionally can induce LV dysfunction, and, very rarely, VF or polymorphic VT. It is supposed to be a relatively benign condition (**Viskin S, et al The "short-coupled" variant of right ventricular outflow ventricular tachycardia: a not-so-benign form of benign ventricular tachycardia? J Cardiovasc Electrophysiol. 2005 Aug; 16(8):912-6**.) Until relatively recently, outflow tract (OFT) arrhythmias OFT PVCs were considered benign. However, this notion has been invalidated by reports over the last 15 years, showing that a small cohort of these patients can present with polymorphic VT/VF (**Haïssaguerre M, Shoda M, Jaïs P, Nogami A, Shah DC, Kautzner J, et al. Mapping and ablation of idiopathic ventricular fibrillation. Circulation 2002; 106:962–7.** (**Haïssaguerre M, Extramiana F, Hocini M, Cauchemez B, Jaïs P, Cabrera JA, et al. Mapping and ablation of ventricular fibrillation associated with Long-QT and Brugada syndromes. Circulation 2003; 108:925–8**.) (**Shimizu W. Arrhythmias origination from the right ventricular outflow tract: how to distinguish "malignant" from "benign"? Heart Rhythm 2009; 6:1507–11.** (**Viskin S, Rosso R, Rogowski O, Belhassen B. The "shortcoupled" variant of right ventricular outflow ventricular tachycardia: a not-so-benign form of benign ventricular tachycardia? J Cardiovasc Electrophysiol 2005; 16:912–6.**). This condition may be mistaken for ACM or idiopathic VF. Since malignant OFT arrhythmias are amenable to definitive cure with RFCA, it is imperative to recognize this variant. The key is to show that a patient's isolated and putatively benign PVCs share the same morphology as the PVCs that trigger the malignant arrhythmia, indicating an identical origin for both phenomena. Successfully targeting the triggering PVC is sufficient to effect cure of polymorphic VT or VF; however, it is advisable to also implant an internal CDI should the PVC suddenly reemerge. In general, malignant OFT triggering PVCs have short coupled, usually landing on the preceding T wave sinus beat. The coupling interval is often but not always shorter than that associated with benign OFT PVCs (and is longer than that associated

with idiopathic VF) (**Viskin S, Rosso R, Rogowski O, Belhassen B. The "shortcoupled" variant of right ventricular outflow ventricular tachycardia: a not-so-benign form of benign ventricular tachycardia? J Cardiovasc Electrophysiol 2005; 16:912–6) (Noda T, et al. Malignant entity of idiopathic ventricular fibrillation and polymorphic ventricular tachycardia initiated by premature extrasystoles origination from the right ventricular outflow tract. J Am Coll Cardiol 2005; 46:1288–94**.). Igarashi et al. suggested that a prematurity index (coupling interval PVC/sinus cycle length) distinguishes between malignant from benign PVCs. A prematurity index r0.73 has a sensitivity of 91% and a specificity of 44% for identifying malignant PVCs (**Igarashi M, Tada H, Kurosaki K, Yamasaki H, Akiyama D, Sekiguchi Y, et al. Electrocardiographic determinants of the polymorphic QRS morphology in idiopathic right ventricular outflow tract tachycardia. J Cardiovasc Electrophysiol 2014; 23:521–6**.). However, Kim et al. not confirmed this finding and have instead proposed that the coupling interval of the second PVC during runs of NSVT better differentiates between malignant and benign forms of OFT PVCs (313 ms vs. 385 ms, respectively) (**Kim YR, et al. Second coupling interval of nonsustained ventricular tachycardia to distinguish malignant from benign outflow tract ventricular tachycardias. Heart Rhythm 2014; 11:2222–30**.). At present, it is reasonable to conclude that the best metric for identifying malignant PVCs is unsettled and that no single parameter consistently distinguishes between malignant and benign OFT PVCs.

XV) Management

- **ACM:** Restriction from high endurance and competitive sports (Exercise has a disproportionate role in the pathogenesis of ACM in patients without desmosomal mutations. (***Sawant AC et al.* Exercise has a disproportionate role in the pathogenesis of arrhythmogenic right ventricular dysplasia/cardiomyopathy in patients without desmosomal mutations *J Am Heart Assoc. 2014 Dec; 3(6):e001471***.) (**Sawant AC, et al. Safety of American Heart Association-**

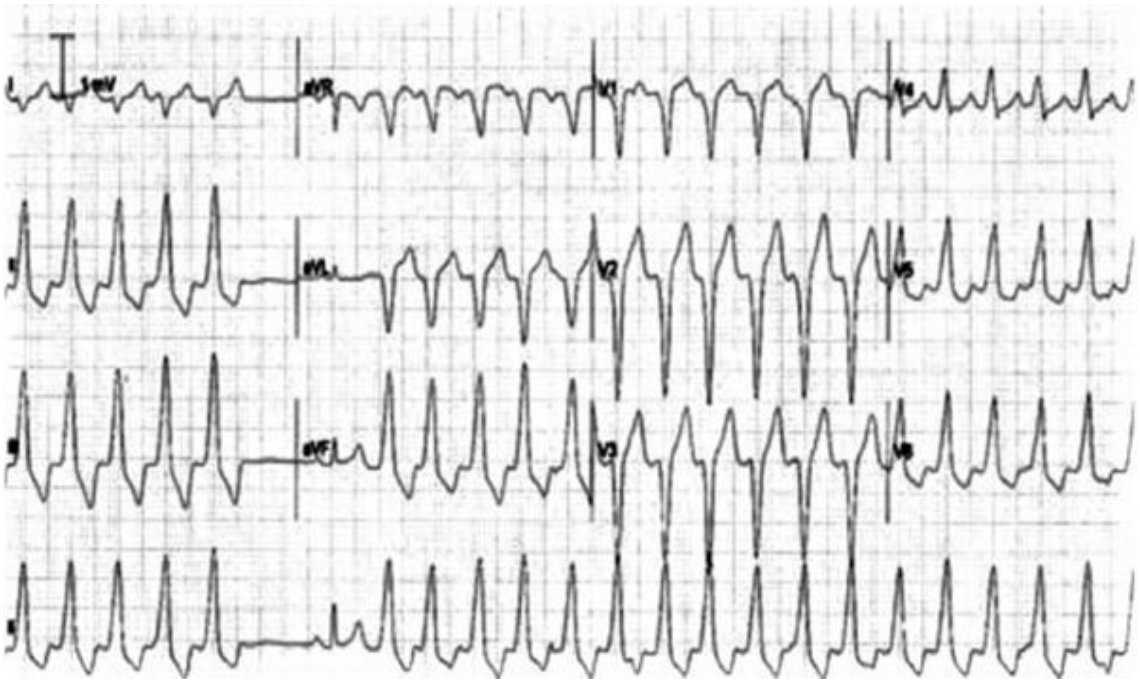
recommended minimum exercise for desmosomal mutation carriers.Heart Rhythm. 2016 Jan; 13(1):199-207.)

B- blockers (BBs): CHF treatment,

ICDs: In cases of S-VT, ventricular flutter (as defined as a CL ≤ 240 msec), or VF

Hemodynamically stable VT (polemic): Combined endocardial/epicardial RFCA of choice for recurrent MACE in ACM. patients with ACM who experience a SVT arrhythmia, regardless of hemodynamic stability, have a sufficiently high risk of SCD to warrant placement of an ICD (**Orgeron GM, et al. Implantable Cardioverter-Defibrillator Therapy in Arrhythmogenic Right Ventricular Dysplasia / Cardiomyopathy: predictors of Appropriate Therapy, Outcomes, and Complications. *J AM Heart Assoc.* 2017: e006242**.)

- **Idiopathic RVOT-VT:** Treatment options include medical therapy vs. RFCA of RVOT. Acute termination of RVOT VT can be achieved by vagal maneuver or adenosine (6 mg up to 24 mg). Intravenous verapamil (10 mg given over 1 min.) is an alternative if the patient has adequate blood pressure. RFCA of RVOT has acute success rates > 80% (range, 85% to 100%). Recurrence up to 5% of cases with the mean recurrence rate of 7%.



12-lead ECG of a 36-year-old pregnant woman admitted with a 4-week history of increasingly intrusive palpitations associated with presyncope. Bursts of broad complex VT are seen with a LBBB morphology, inferior axis (right) with tall R waves in leads II, III, and aVF and precordial transition at V4 consistent with origin from the RVOT.
From **Hogarth AJ, Graham LN. Normal heart ventricular tachycardia associated with pregnancy: successful treatment with catheter ablation. Indian Pacing Electrophysiol J March 12, 2014;14(2):79–8**2 with permission.

A scoring system was proposed by Hoffmayer et al (**Hoffmayer KS, Bhave PD, Marcus GM, *et al.* An electrocardiographic scoring system for distinguishing right ventricular outflow tract arrhythmias in patients with arrhythmogenic right ventricular cardiomyopathy from idiopathic ventricular tachycardia. Heart Rhythm. 2013; 10:477-82. doi: 10.1016/j.hrthm.2012.12.009**) to distinguish between ACM from idiopathic VT provides the following values:

- 3 points for sinus rhythm anterior T-wave inversions in leads V1–V3 and during ventricular arrhythmia;
- 2 points for QRS duration in lead I ≥120 ms;
- 2 points for QRS notching;
- 1 point for precordial transitional lead V5 or later.

A score of 5 or greater was able to correctly distinguish ACM from idiopathic VT 93% of the time, with a sensitivity of 84%, specificity of 100%, positive predictive value of 100%, and negative predictive value of 91%.