

# ECG - Checklists for Sports Cardiology Evaluation

(Löllgen, Houston, 2018)

## Cardiomyopathies

- HCM, NC CMP, DCM
  - ARVD

## Marfansyndrome

- Electrical diseases
- Long-QT Syndrom
- BrugadaSyndrom
  - CPVT

# Why Checklists: Young Males ECG: Six percent with abnormal ECG Reduce Sudden death !! (Seattle Criteria)

	Nbr of Detections	Percentage [%]
Total number of analysed resting ECGs	274'468	100.00
Abnormal ECG finding	17'765	6.47
T-wave inversion	1'193	0.43
ST segment depression	3'080	1.12
Pathologic Q waves	5'947	2.17
Complete left bundle branch block	92	0.03
Intraventricular conduction delay	385	0.14
Left axis deviation	2'800	1.02
Left atrial enlargement	1'709	0.62
Right ventricular hypertrophy pattern	991	0.36
Ventricular pre-excitation	71	0.03
Long QT interval	1'412	0.51
Short QT interval	6	0.00
Brugada-like ECG pattern	0	0.00
Profound sinus bradycardia	0	0.00
Atrial tachyarrhythmias	1'599	0.58
Premature ventricular contractions	416	0.15
Ventricular arrhythmias	126	0.05
Other abnormality	162	0.06

Table 2 Absolute and relative numbers of Seattle Criteria detections.

(N= 440000, Swiss Army)

(Abaecherli et al., 2014)

H. Löligen, EFMD 2019

# Goals of Examination:

## Detection of Occult Diseases (Priori,2013,2015 )

### >>> Structural („electrical“) abnormalities (Priori,2014)

- **Ion channel diseases:** long and short QT-syndrome, Brugada-syndrome, early repolarisation
- **Preexcitation Syndrom** (WPW) with intermitt.atrial tachycardia and esp. atrial fibrillation),
- **CPVT** Catecholaminergic polymorphic ventricular tachykardia
- Progressive **cardiac conduction disease**, unexplained cardiac arrest (idiopathic VF)
- **Unexplained cardiac arrest:** Sudden unexplained death syndrom and sudden unexplained death in infancy
- >>> **Acquired diseases : most causes in elderly**  
**Coronary artery disease**, arterial hypertension, drugs (TdP), myocarditis, contusio cordis, concussion

# Structural Abnormalities of the Heart

## > **Structural abnormalities** (Heart muscle)

- **Cardiomyopathies:** hypertrophic \*, with or without outflow obstruction, dilatative, non- compaction cm, arrhythmogen (ARVD), (**„Moges**)
  - Marfan-Syndrome,
  - Abnormal findings more in Afro-Americans and in Basketball players  
(e.g. Marfan-Syndrom) (ECG abnormal in 3.7 %,pot. lethal 0.3%, (Asif, BJSM 2014)
- CAD, Coronary abnormalities

\*n =750000 with HCM in US, diagnosed in 100000 of them,  
mostly non-invasive imaging (Maron, NEJM, Aug. 2018)



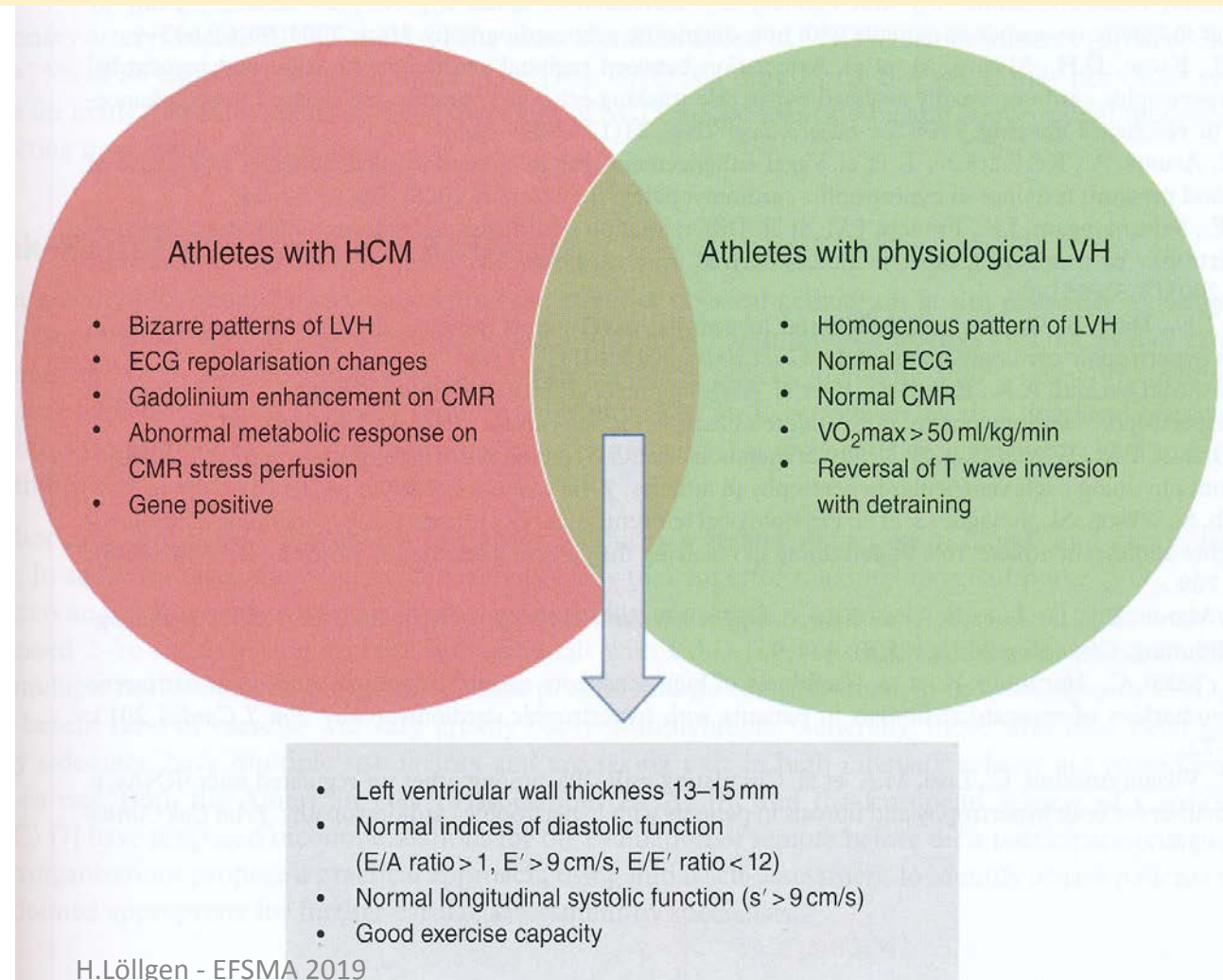
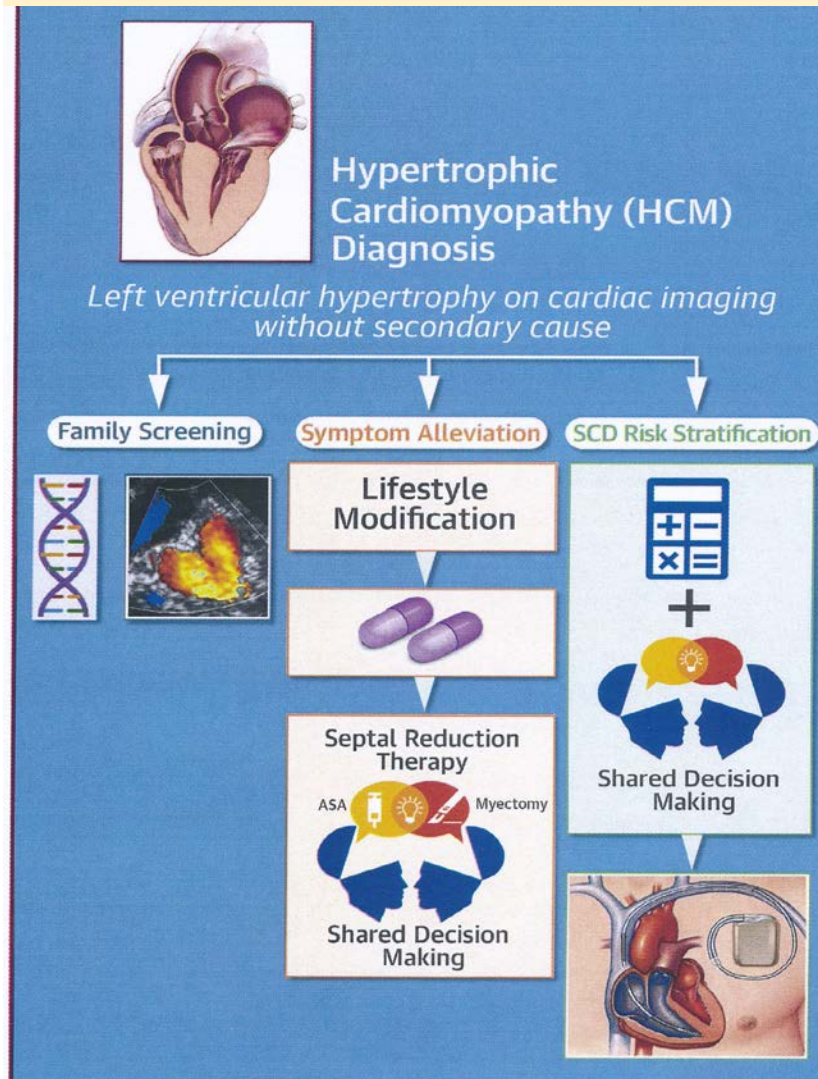
# „Electrical“ causes of cardiac problems in athletes

Ion- Channel diseases or congenital arrhythmogenic diseases

- Long - QT-Syndrome (LQT1 – 8) :  
congenital, acquired (drugs)  
(z.B. Jervell-Lange-Nielsen-Syndrom) (JLNS 1 – 2)
  - Short QT-Syndrome (SQTS 1 -3)  
Brugada –Syndrome (BrS 1 – 3)
  - Catecholaminergic polymorph ventricular tachycardia  
(CPVT 1- 2)
- Conduction disturbances

# HCM Diagnosis

Geske et al., JACC HF, 2018



# HCM : Checklist

Prinz et al., Dtsch Ärztebl Int 2011

## Algorithm to identify at-risk HCM patients for implantation with an ICD 25

First degree risk factors	Definition
Positive family history of sudden cardiac death	Cases with SCD <45 years
Recurrent syncope	$\geq 2$ incidents
LVH	$\geq 30$ mm at any site in the LV
Abnormal blood pressure response during exercise	Increase <20 mm hg or fall >20 mm Hg after transient increase
Non-sustained VT in Holter ECG	$\geq 3$ consecutive QRS complexes with a heart rate of $\geq 120$ bpm.
<b>Second degree risk factors</b>	
Atrial fibrillations/atrial flutter LA dilatation High LVOT gradient at rest	Any form, provided cannot be eliminated >45 mm (in m-mode ECG)
Evidence of myocardial ischemia during exercise	>80 mm Hg (CW Doppler)
Early manifestation of HCM Myocardial bridging near the LAD Marked fibrosis in cardiac MRI	<30 years of age in younger patients (<45 years) fibrosis of $\geq 2$ segments in a 17-segment model of the LV

SCD, sudden cardiac death; LVH, left ventricular hypertrophy; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; VT, ventricular tachycardia; LAD, left anterior descending artery



# Moge(s) Nosology System for Classifying CM Patients (Arbustini, 2016)

NOTATION	M MORPHO-FUNCTIONAL PHENOTYPE	O ORGAN/SYSTEM INVOLVEMENT	G GENETIC INHERITANCE PATTERN	E ETIOLOGY	S STAGE
CHARACTERISTICS	<p>Proband's cardiomyopathy (CM) diagnosis (DCM, HCM, RCM, ARVC/D, LVNC)</p>	<p>Clinical history and evaluation</p> <ul style="list-style-type: none"> <li>Organ involvement: Extracardiac organs/tissues</li> <li>Multidisciplinary evaluation according per clinical needs or diagnostic hypothesis</li> </ul>	<p>Genetic counseling with pedigree</p> <ul style="list-style-type: none"> <li>Familial                             <ul style="list-style-type: none"> <li>Inheritance AD, AR XL (R or D) or Matrilineal</li> </ul> </li> <li>Non-familial; Phenotypically sporadic                             <ul style="list-style-type: none"> <li>Informative and non-informative families</li> <li>Consultant non-informed about family history</li> </ul> </li> </ul> <p>Clinical family screening</p> <ul style="list-style-type: none"> <li>Affected, asymptomatic relative unaware of the disease</li> <li>Relatives with ECG and/or Echo abnormalities</li> <li>Healthy family members with normal ECG and ECHO</li> </ul>	<p>Genetic testing in the proband</p> <ul style="list-style-type: none"> <li>Positive                             <ul style="list-style-type: none"> <li>Cascade genetic testing in relatives</li> </ul> </li> <li>Negative                             <ul style="list-style-type: none"> <li>New tests novel genes</li> <li>Regular monitoring in relatives</li> </ul> </li> </ul>	<p>Functional status ACC/AHA, NYHA</p>
SUBSCRIPT	<p>D Dilated</p> <p>H Hypertrophic</p> <p>R Restrictive</p> <p>R EMF Endomyocardial fibrosis LV=left ventricle RV=right ventricle RLV=biventricular</p> <p>A ARVC M=major m=minor c=category LV= left ventricle RV=right ventricle RLV=biventricular</p> <p>NC LVNC</p> <p>E Early, with type in parentheses</p> <p>NS Nonspecific phenotype</p> <p>NA Information non available</p> <p>O Unaffected*</p>	<p>H Heart LV=left ventricle RV=right ventricle RLV=biventricular</p> <p>M Muscle (skeletal)</p> <p>N Nervous</p> <p>C Cutaneous</p> <p>E Eye, Ocular</p> <p>A Auditory</p> <p>K Kidney</p> <p>G Gastrointestinal</p> <p>Li Liver</p> <p>Lu Lung</p> <p>S Skeletal</p> <p>O Absence of organ/system involvement*, e.g. in family members who are healthy mutation carriers; the mutation is specified in E and inheritance in G</p>	<p>N Family history negative</p> <p>U Family history unknown</p> <p>AD Autosomal dominant</p> <p>AR Autosomal recessive</p> <p>XLD X-linked dominant</p> <p>XLR X-linked recessive</p> <p>XL X-linked</p> <p>M Matrilineal</p> <p>O Family history not investigated*</p> <p>Undet Inheritance still undetermined</p> <p>S Phenotypically Sporadic (apparent or real)</p>	<p>G Genetic cause</p> <p>OC Obligate carrier</p> <p>ONC Obligate non-carrier</p> <p>DN De novo</p> <p>Neg Genetic test negative for the known familial mutation</p> <p>N Genetic defect not identified</p> <p>O No genetic test, any reason*</p> <p>G-A-TTR Genetic amyloidosis</p> <p>G-HFE Hemochromatosis</p> <p>Non-genetic etiologies:</p> <p>M Myocarditis</p> <p>V Viral infection (add the virus identified in affected heart)</p> <p>AI Autoimmune/immune-mediate; suspected (AI-S), proven (AI-P)</p> <p>A Amyloidosis (add type: A-K, A-L, A-SAA)</p> <p>I Infectious, non viral (add the infectious agent)</p> <p>T Toxicity (add cause/drug)</p> <p>Eo Hypereosinophilic heart disease</p>	<p>ACC-AHA stage represented as letter A, B, C, D</p> <p>NA not applicable</p> <p>NU not used</p> <p>followed by NYHA class represented as Roman numeral I, II, III, IV</p>



# ECG in Athletes vs. HOCM

Rachel.E.Bent et al., JACC 2015

**TABLE 1 Optimal Electrocardiography Characteristics**

	Points	Hypertrophic Cardiomyopathy		Athlete		Multivariate Regression	
		% With Characteristic	% With Isolated Characteristic	% With Characteristic	% With Isolated Characteristic	Odds Ratio	p Value
TWI <0 mV in V <sub>4</sub> , V <sub>5</sub> , V <sub>6</sub>	1	37.9	8.3	0.80	0.71	87.1	<0.001
TWI < -0.5 mV in V <sub>4</sub> , V <sub>5</sub> , V <sub>6</sub> (major)	2	12.1	1.3	0	0		
QTc >480 ms	1	14.6	1.7	0.44	0.44	3.9	0.41
Q-wave >40 ms in V <sub>5</sub> or aVF	1	6.3	2.1	0.18	0.18	4.6	0.40
ST-segment depression <-0.05 mV V <sub>5</sub> or V <sub>6</sub>	1	33.3	4.2	0.27	0.18	13.4	0.033
ST-segment depression <-0.1 mV V <sub>5</sub> or V <sub>6</sub> (major)	2	15.4	0.83	0	0		
Left atrial abnormality	1	14.2	2.5	1.2	1.2	4.2	0.13
Left axis deviation	1	16.3	5.0	0.80	0.80	22.9	0.003
Right axis deviation	1	4.2	2.5	0.36	0.36	17.0	0.025
QRS duration >140 ms	1	12.5	2.5	0.089	0.089	9.3	0.18
Left bundle branch block (major)	2	5.9	0.83	0	0		

The multivariate regression model was adjusted for age, sex, ethnicity, heart rate, and all of the other electrocardiography characteristics in the first column.

TWI = T-wave inversion.

In support „of the digital ECG“ in athletic screening“

# Checklist: Criteria for ARVD: Major and Minor

S.Priori et al., ESC Guideline, 2015

## Diagnostic criteria[edit]

There is no pathognomonic feature of ARVD. The diagnosis of ARVD is based on a combination of major and minor criteria. To make a diagnosis of ARVD requires either 2 major criteria or 1 major and 2 minor criteria or 4 minor criteria.

### Major criteria

- Right ventricular dysfunction
- Severe dilatation and reduction of RV ejection fraction with little or no LV impairment
- Localized RV aneurysms
- Severe segmental dilatation of the RV
- Tissue characterization
- Fibrofatty replacement of myocardium on endomyocardial biopsy
- Conduction abnormalities
- Epsilon waves in V<sub>1</sub> – V<sub>3</sub>
- Localized prolongation (>110 ms) of QRS in V<sub>1</sub> – V<sub>3</sub>
- Family history
- Familial disease confirmed on autopsy or surgery

### Minor criteria

- Right ventricular dysfunction
- Mild global RV dilatation and/or reduced ejection fraction with normal LV.
- Mild segmental dilatation of the RV
- Regional RV hypokinesis
- Tissue characterization
- Conduction abnormalities
- Inverted T waves in V<sub>2</sub> and V<sub>3</sub> in an individual over 12 years old, in the absence of a right bundle branch block (RBBB)
- Late potentials on signal averaged EKG.
- Ventricular tachycardia with a left bundle branch block (LBBB) morphology
- Frequent PVCs (> 1000 PVCs / 24 hours)
- Family history
- Family history of sudden cardiac death before age 35
- Family history of ARVD



# In Detail : Task Force Criteria for ARVD

## Major and Minor Criteria

(NEJM,2017, D'Ascenzi et al., JACC,2018)

**Table 1. International Task Force Criteria for the Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy.\***

Category	Major Criteria	Minor Criteria
Global or regional dysfunction and structural alteration†		
On two-dimensional echocardiography	Regional RV akinesia, dyskinesia, or aneurysm and one of the following (end diastole): PLAX RVOT $\geq 32$ mm ( $\geq 19$ mm per square meter when corrected for body-surface area), PSAX RVOT $\geq 36$ mm ( $\geq 21$ mm per square meter when corrected for body-surface area), or fractional area change of $\leq 33\%$	Regional RV akinesia or dyskinesia and one of the following (end diastole): PLAX RVOT 29 to $<32$ mm (16 to $<19$ mm per square meter when corrected for body-surface area), PSAX RVOT 32 to $<36$ mm (18 to $<21$ mm per square meter when corrected for body-surface area), or fractional area change of 34 to 40%
On MRI	Regional RV akinesia or dyskinesia or dyssynchronous RV contraction and one of the following: ratio of RV end-diastolic volume to body-surface area $\geq 110$ ml per square meter (male patients) or $\geq 100$ ml per square meter (female patients), or RV ejection fraction $\leq 40\%$	Regional RV akinesia or dyskinesia or dyssynchronous RV contraction and one of the following: ratio of RV end-diastolic volume to body-surface area 100 to $<110$ ml per square meter (male patients) or 90 to $<100$ ml per square meter (female patients), or RV ejection fraction 41 to 45%
On RV angiography	Regional RV akinesia, dyskinesia, or aneurysm	
Tissue characterization	$<60\%$ residual myocytes on morphometric analysis (or $<50\%$ , if estimated) and fibrous replacement of the RV free-wall myocardium, with or without fatty replacement of tissue, in at least one endomyocardial-biopsy sample	60 to 75% residual myocytes, on morphometric analysis (or 50 to 65%, if estimated) and fibrous replacement of the RV free-wall myocardium, with or without fatty replacement of tissue, in at least one endomyocardial-biopsy sample
Repolarization abnormalities	Inverted T waves in right precordial leads ( $V_1$ , $V_2$ , and $V_3$ ) or beyond in patients older than 14 yr of age (in the absence of complete right bundle-branch block, QRS $\geq 120$ msec)	Inverted T waves in leads $V_1$ and $V_2$ in patients older than 14 yr of age (in the absence of complete right bundle-branch block) or in $V_4$ , $V_5$ , or $V_6$ ; inverted T waves in leads $V_1$ , $V_2$ , $V_3$ , and $V_4$ in patients older than 14 yr of age (in the presence of complete right bundle-branch block)
Depolarization and conduction abnormalities	Epsilon wave (reproducible low-amplitude signals from end of QRS complex to onset of the T wave) in the right precordial leads ( $V_1$ , $V_2$ , and $V_3$ )	Late potentials on signal-averaged ECG in at least one of three parameters in the absence of a QRS complex duration of $\geq 110$ msec on the standard ECG; filtered QRS complex duration, $\geq 114$ msec; duration of terminal QRS complex $<40$ $\mu$ V (low-amplitude signal duration), $\geq 38$ msec; root-mean-square voltage of terminal 40 msec, $\leq 20$ $\mu$ V; terminal activation duration of QRS complex, $\geq 55$ msec, measured from the nadir of the S wave to the end of the QRS complex, including R', in $V_1$ , $V_2$ , or $V_3$ , in the absence of complete right bundle-branch block
Arrhythmias	Nonsustained or sustained ventricular tachycardia with a left bundle-branch block and superior axis pattern (negative or indeterminate QRS complex in leads II, III, and aVF and positive QRS complex in lead aVL)	Nonsustained or sustained ventricular tachycardia of RV outflow configuration with a left bundle-branch block and inferior axis pattern (positive QRS complex in leads II, III, and aVF and negative QRS complex in lead aVL) or unknown axis, or $>500$ ventricular extrasystoles per 24 hr (on Holter monitoring)
Family history	ARVC confirmed in a first-degree relative who meets current task-force criteria, ARVC confirmed pathologically at autopsy or surgery in a first-degree relative, or identification of a pathogenic mutation categorized as associated or probably associated with ARVC in the patient under evaluation‡	History of ARVC in a first-degree relative in whom it is not possible or practical to determine whether current task-force criteria are met, premature sudden death (at $<35$ yr of age) due to suspected ARVC in a first-degree relative, or ARVC confirmed pathologically or by current task-force criteria in a second-degree relative

\* The table is adapted from Marcus et al.<sup>49</sup> The diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC) is considered to be definite if the patient meets two major criteria, one major and two minor criteria, or four minor criteria from different categories; the diagnosis is considered to be borderline if the patient meets one major and one minor criteria or three minor criteria from different categories, and the diagnosis is classified as possible if the patient meets one major or two minor criteria from different categories. ECG denotes electrocardiogram, PLAX parasternal long-axis view, PSAX parasternal short-axis view, RV right ventricular, and RVOT RV outflow tract.

† Hypokinesia is not included in this or subsequent definitions of RV regional wall-motion abnormalities for the proposed modified criteria.

‡ A pathogenic mutation is a DNA alteration associated with ARVC that alters or is expected to alter the encoded protein, is unobserved or rare in a large, non-ARVC control population, and either alters or is predicted to alter the structure or function of the protein or has shown linkage to the disease phenotype in a conclusive pedigree (i.e., a pedigree providing conclusive evidence of a mendelian inheritance of the disease phenotype).



# ARVD: Clinical Indicators ... Differentiation between Athletic Remodeling and ARVD (Zaidi et al., JACC 2015)

