

# Update on Cardiac Imaging in Syncope

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SJH

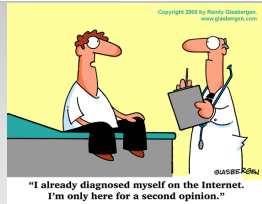
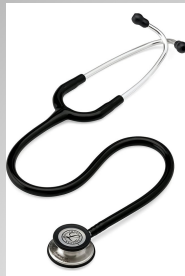
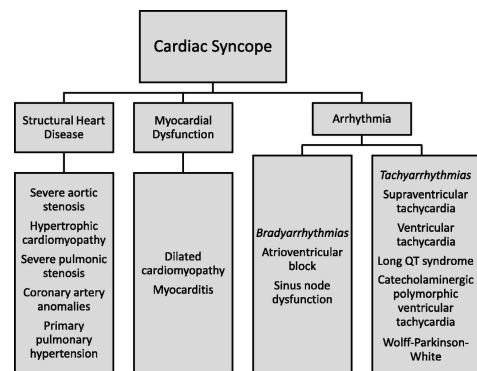
- NONCARDIAC**
- Neurologic**
    - Seizure disorder
    - Narcolepsy
    - Hydrocephalus
    - Subarachnoid hemorrhage
    - Migraine
  - Cerebrovascular Disease**
    - Vertebrobasilar disease/subclavian steal
    - Carotid artery disease
  - Metabolic/Endocrine Abnormality**
    - Hypo/hyperglycemia
    - Hypoxia
    - Volume depletion
    - Hypocapnia
    - Systemic mastocytosis, carcinoid
  - Psychiatric Disturbances**
    - Hysteria
    - Panic attacks
- CARDIAC**
- Structural Cardiovascular Abnormality**
    - Myocardial disease/ischemia
    - Pericardial disease/valvular disease
    - Hypertrophic obstructive cardiomyopathy
    - Cardiac tumors
    - Pulmonary hypertension/embolus
  - Primary Cardiac Rhythm Disturbance**
    - Ischemic/hemodynamic
    - Brady/tachyarrhythmia
  - Neurally-Mediated Disturbances of Blood Pressure Control**
    - Vasovagal syncope
    - Respiratory/nausea syncope
    - Airway stimulation
    - Gastrointestinal-deglutition
    - Defecation syncope
    - Urologic-micturition syncope
    - Carotid sinus hypersensitivity
  - Dysautonomic/Orthostatic Disturbances of Blood Pressure Control**
    - Primary autonomic failure
    - Diabetic neuropathy/amyloidosis
    - Drug induced orthostasis

Etiology of syncope	Adjusted risk of death
Cardiac	2.01*
Neurologic	1.54*
Idiopathic	1.32*
Vasovagal	1.08

NEJM 2002;347:878

\*p<0.01

**Prognosis : Framingham 25 year follow up**



**Clinical examination and ECG**

## Cardiac Structural

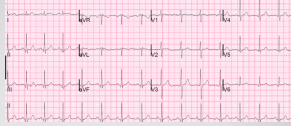
- Exertional
- Chest pain or SOB
- Abnormal Exam
- Murmurs \*\*\*\*\*
- Abnormal ECG

## Cardiac Arrhythmia

- Arrhythmia in "normal" heart or structurally abnormal
- May not be exertional – eg bradycardia
- Supine syncope scary
- Palpitations
- ECG absolutely key – but may be normal

**Clinical decision making**

- 24 year old male
- Football x 3 weekly
- Brought in by ambulance after collapse on playing field
- Chest discomfort at peak exertion
- Normal clinical examination
- Normal echocardiogram



## Exertional Syncope



## MRA of malignant course of RCA

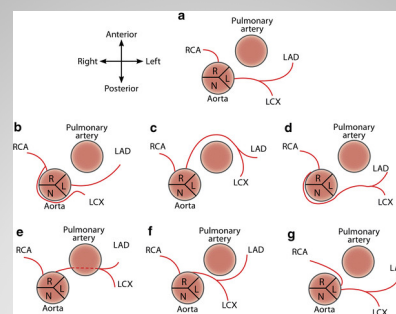
1496 Basso et al. **Coronary Anomalies and Sudden Death in Athletes** JACC Vol. 35, No. 6, 2000 May 2000:1493-501

**Table 2.** Demographic and Clinical Data in 12 Athletes With Wrong Sinus Coronary Artery Origin and Clinical Manifestations or Diagnostic Testing During Life

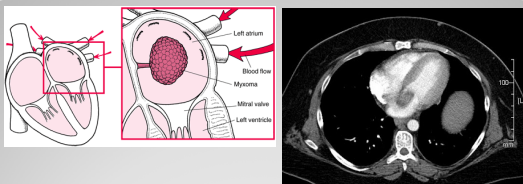
Age at Death	Gender	Nation	Race	Sport	Level	Site	Activity	Circumstances of Death	Prior Symptoms	Time From Symptoms to Sudden Death
11	M	Italy	W	Soccer	JHS	Field	Game	During effort	No	—
12	M	U.S.	B	Basketball	JHS	Field	Practice	During effort	Chest pain	7 days
12	M	U.S.	W	Hockey	JHS	Hond	Sedentary	After effort	Syncope*, chest pain*	15 mo
12	M	U.S.	B	Basketball	JHS	Field	Practice	During effort	Syncope*†	14 mo
14	M	Italy	W	Soccer	JHS	Field	Game	During effort	No	—
15	M	Italy	W	Soccer	JHS	Field	Game	During effort	Syncope*	11 mo
15	F	U.S.	W	Trackpoint	HS	Field	Practice	During effort	Dizziness, palpitations*	15 mo
15	M	U.S.	B	Basketball	JHS	Field	Practice	During effort	Syncope†, chest pain	24 mo
16	M	U.S.	B	Basketball	HS	Field	Game	During effort	Chest pain*	8 mo
22	M	Italy	W	Soccer	Pro	Field	Game	During effort	Palpitations	12 mo
29	M	Italy	W	Rugby	Pro	Field	Practice	During effort	Palpitations	13 mo
32	F	Italy	W	Running	Pro	Field	Game	During effort	Chest pain*	9 mo

\*During physical exertion; †recurrent episodes; coronary artery anatomy was not investigated; — = data not available. JHS = high school; HS = junior high school; LMSA = left main coronary artery origin from left aortic sinus; mo = months; pro = professional; PFC = prefrontal cortex; ventricular complex; RCA = right coronary artery origin from right aortic sinus; U.S. = United States.

## Syncope and anomalous coronary



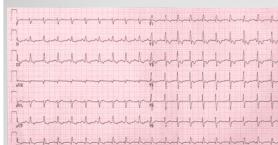
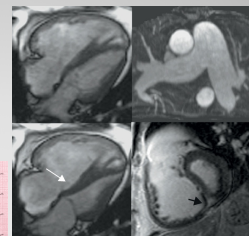
## Anomalous coronary arteries



- Myxoma or other cardiac tumours
- Pulmonary hypertension
- Outflow tract obstruction
  - Aortic Stenosis
  - HCM
  - Sub-aortic membrane

## Direct reduction in Cardiac Output

- 21 year old female
- Student nurse
- Collapse x 2
- On climbing stairs
- Mild SOB



### Syncope and NYHA FC at time of diagnosis in registries

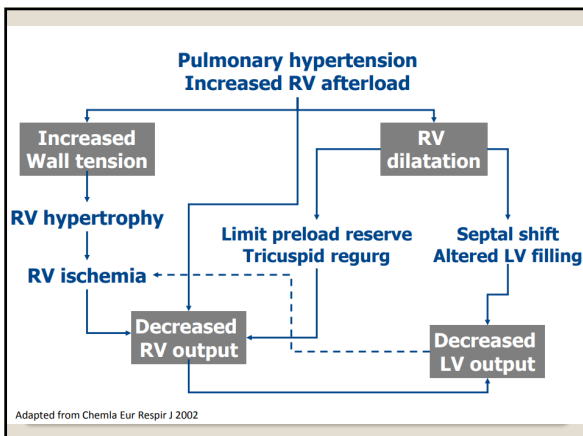
	Rich Ann intern Med 1987	Thenappan ERJ 2007	Tueller Swiss M W 2008	Humbert AJRCCM 2006	Jin CHEST 2007	Benza Circulation 2010
Year	1981-1985	1982-2006	1999-2008	2002-2003	2005	2006-2007
Distribution	National (USA)	1 centre (Chicago)	National (Swiss)	National (France)	3 centres (China)	National (USA)
Patients, n	187	245	192	674	72	2716
Syncope, %	13	-	-	-	26	-(12*)
WHO FC, %						
II	29	20	12	24	37	37.8
III			62	63	44	48.2
IV	71 III/IV	80 III/IV	25	12	17	5.5

> 10 % of patients with PAH present syncope at diagnosis

### Syncope and near-syncope in the NYHA FC

#### Level of physical activity carried out by an individual that does not lead to symptoms (or signs of RV failure)

- Patients with PH without limitation of physical activity. Ordinary activity does not cause undue dyspnoea or fatigue, chest pain or **near syncope**
- Patients with PH resulting in slight limitation of physical activity. Comfortable at rest. Ordinary physical activity causes undue dyspnoea or fatigue, chest pain or **near syncope**
- Patients with PH resulting in marked limitation. Less than ordinary activity causes undue fatigue or dyspnoea, chest pain or **near syncope**
- Patients with PH with inability to carry out any physical activity. Signs of right heart failure. **Syncope\***



#### Loss of ventricular adaptation to afterload

- Decreased RV output and RV/LV interdependence
- Uncoupling between RV and pulmonary circulation
- Unmet demand to increase CO during exercise
- Drop in systemic pressure due to abrupt decrease in SV

#### Other (uncommon) contributing factors

- Arrhythmias
- RV and/or LV ischemia
  - Subendocardial microcirculation
  - Dynamic occlusion of LAD due to PA dilatation
- Severe PA dilatation

- 31 year old man
- Working on fish farm in Donegal
- Collapse and fell into water
- Rescued by co worker-Letterkenny
- Multiple episodes of VT in CCU
- Interventricular conduction delay on ECG
- Normal ECHO
- Father RIP suddenly age 47



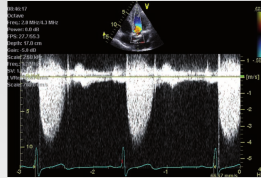
### Recurrent VT

- Conduction abnormality
  - CHB 23-30%
  - First degree avb, bbb
- Arrhythmias
  - VT 23%
  - AF/Flutter 15-23%
- SCD
  - Terminal event 67%
- CHF
  - Progressive 25%
- Other
  - Pericarditis/effusion, constriction (3-19%), tamponade rarely

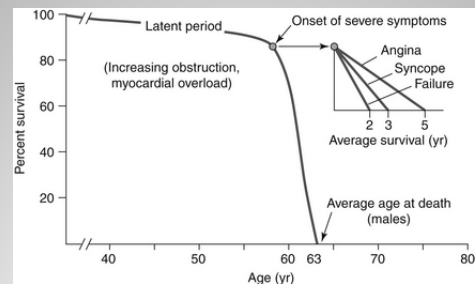


### Cardiac Sarcoid

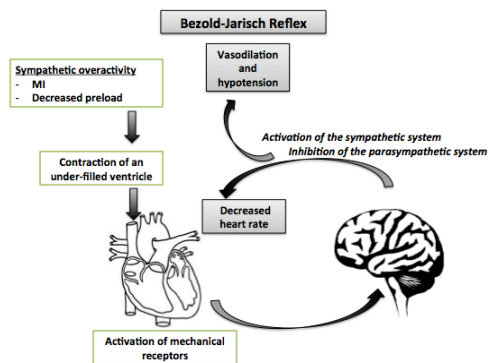
- 54 year old man
- Syncope on holiday in Spain in context of > usual alcohol
- Cycles to work (3 miles) for 15 years
- Past 3 months SOB and has had to stop 1-2 times on trip
  - Ex smoker
  - BMI 29
  - Chol 7
  - BP 180/90
  - Father RIP age 57 MI



**About to get on the treadmill**

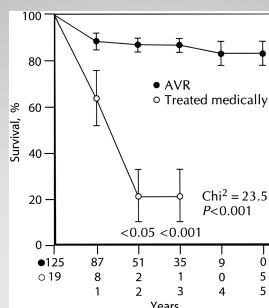


**Natural history Aortic Stenosis**

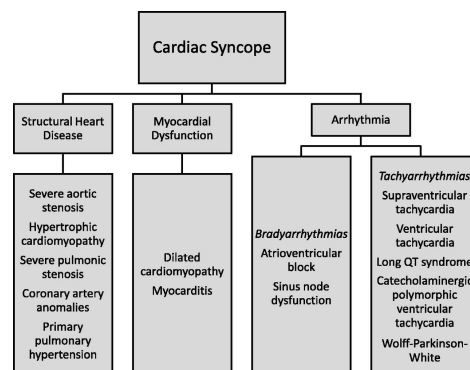


- The often repeated story
  - Excessive stimulation of mechanoreceptors due to forceful contraction of underfilled left ventricle leading to paradoxical signals to the CNS – causing change from vasoconstriction to vasodilation – causing drop in blood pressure and bradycardia
- The more complicated story
  - Disordered baroreflex function, paradoxical cerebral autoregulation, endogenous vasodilators... we don't really know....

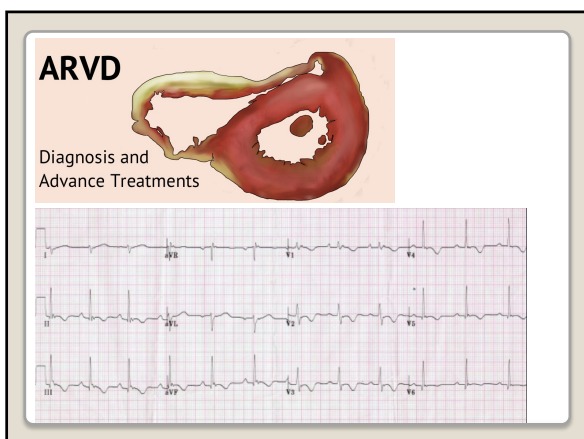
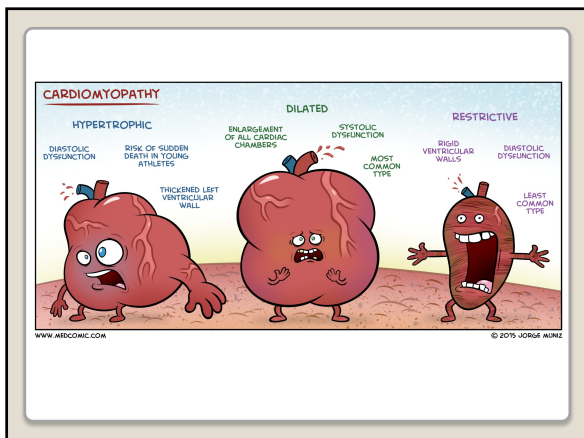
**Bezold-Jarisch phenomenon**



**Corrected survival with AVR**







**ARVD Genes**

Architectural unit of the desmosome

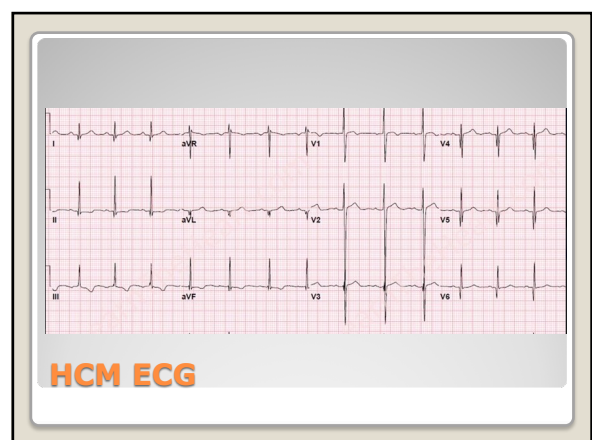
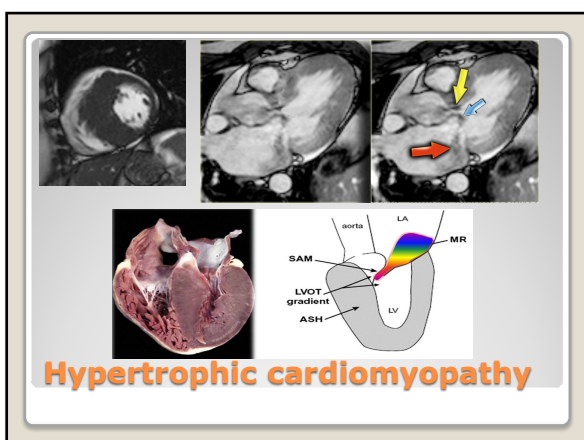
The diagram shows the architectural unit of the desmosome, which is a complex of proteins that anchor intermediate filaments to the cell membrane. The proteins involved are Desmoplakin, Plakophilin, and Desmoglein. The diagram also shows the associated proteins, including Plakoglobin, Desmoplakin 2 (DSP2), and Desmoglein 2 (DSG2).

The responsible genes include:

- Plakophilin 1 (PKP1)
- Desmoplakin 1 (DSP1)
- Plakophilin 2 (PKP2)
- Desmoplakin 2 (DSP2)
- Desmoglein 2 (DSG2) and others

- 50% have family history
- Prevalence 1 in 5000 ( up to 1 in 2000)
- Typically AD, but AR also described
- Clinical course more malignant in men

**ARVD**



#### Mechanisms for syncope in hypertrophic cardiomyopathy

##### Arrhythmia

Paroxysmal atrial fibrillation/supraventricular tachycardia

Complete heart block/sinus node dysfunction

Sustained ventricular tachycardia

Primary haemodynamic mechanism

Left ventricular outflow tract obstruction

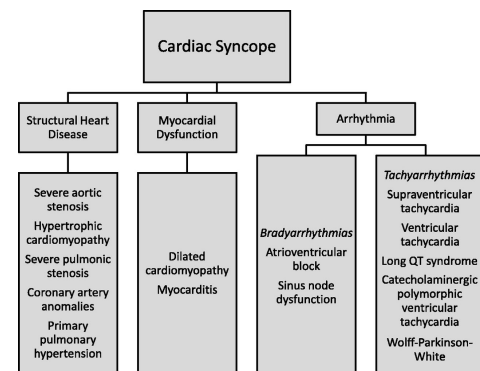
Abnormal vascular control mechanisms leading to episodes of hypotension due to inappropriate vasodilatation

Hypotension due to impaired filling when preload is reduced in the setting of diastolic dysfunction

- Syncope or presyncope in 15-25% of HCM
- Young patients with recurrent syncope at increased risk of SCD
- Prior cardiac arrest or sust. VT- for ICD
- Risk Calculators now available
- Extent of scar on CMR predictive of events\*
- Two or more risk factors for SCD-high risk
  - **Max wall thickness >30**
  - **Fall in BP during exercise**
  - **NSVT on 48 hour holter**
  - **Family HX SCD <45 yrs**
  - **Resting LVOT gradient**
  - \*CMR predictive but difficult to prove independent

#### Hypertrophic cardiomyopathy

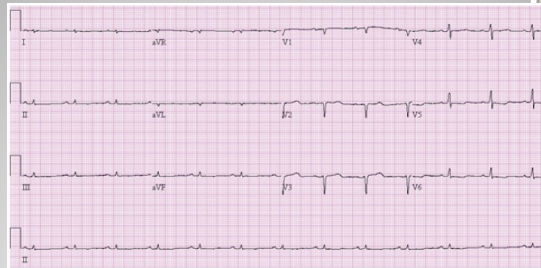
<http://www.doc2do.com/hcm/webHCM.html>



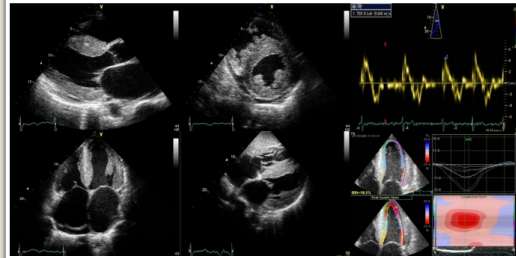
- Tachyarrhythmia
  - SVT – generally better tolerated , but less well so with progressive impairment in Fxn
  - VT – often scar related
- Bradyarrhythmia
  - AV conduction delay (Lamin AC CM)
  - Sinus node disease
- Hypotension
  - Poor CO
  - Vasoactive medication or diuretics

#### Causes of Syncope in HF

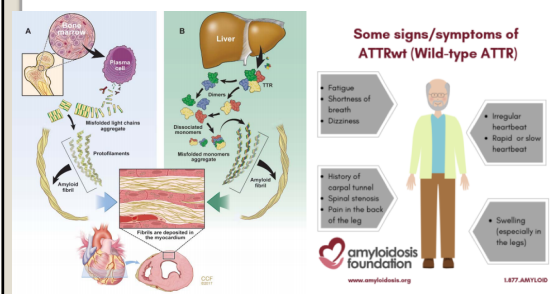
#### Update on imaging



**76 yrs man with syncope and OH**



**Echo of pt with syncope and OH**



#### DIAGNOSIS OF VARIANT TTR CARDIAC AMYLOIDOSIS

1. Clinical suspicion and recognition!
2. Tissue biopsy to identify amyloid by Congo red stain
  - Abdominal fat aspirate (20-25% sensitive for ATTRwt, 50-75% sensitive for ATTRvariant)
  - Endomyocardial biopsy (virtually 100% sensitive/specific)
3. Typing of amyloid by analysis of biopsy specimen
  - Immunohistochemistry (problematic high background)
  - Mass spectrometry (Mayo Medical Labs send out)
4. Genetic testing to establish TTR genotype
  - Blood test - PCR
5. Exclusion of plasma cell dyscrasia

Copyright: Amyloidosis.org

Mayo Medical Labs  
School of Medicine



November 2016

# **Multicenter Study of Planar Technetium 99m Pyrophosphate Cardiac Imaging Predicting Survival for Patients With ATTR Cardiac Amyloidosis**

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<sup>1</sup> Author Affiliations | Article Information

JAMA Cardiol. 2016;2(8):680-689. doi:10.1001/jamacardio.2016.2839

## Multicenter Analysis

	No. of Patients		
	ATTR <sup>+</sup>	ATTR <sup>-</sup>	Total
Positive scan	110	4	114
Negative scan	11	46	57
Total	121	50	171

Sensitivity = 91%

Specificity = 92%

AUC, 0.960 (95% CI, 0.930-0.981)

- Structural abnormalities causing fixed or restricted cardiac output
  - AS, Myxoma, LVOT obstruction, PHT
- Other structural abnormalities
  - Eg Anomalous Coronary Artery
- Myocarditis
  - either through scar and propensity to malignant arrhythmia or reduced CO
- Functional abnormalities
  - DCM/HCM/ARVD– either through scar and propensity to malignant arrhythmia or reduced CO
  - Unusual causes of HCM or HF PEF

**Cardiac Imaging identifies...**