



# What Causes Sudden Cardiac Death

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# Definition Sudden Cardiac Death

- Unexpected death from definite or probable cardiac cause within 1 hour of symptom onset

or

- Unexpected death from probable cardiac cause within 24 hours of last sighting in good health



# Incidence of SCD

- Varies across age groups
- 36-64 years
  - 10 to 75 per 100,000 persons / year
  - > 5000 deaths per year (Ireland)
  - 100,000 deaths per year (UK)
  - 300,000 cardiac arrests attended by EMS in USA
    - May only account for 60% of SCD
- 1-35 years
  - 1 to 3 per 100,000 persons / year
  - 60-80 deaths per year (Ireland) ,400 in UK

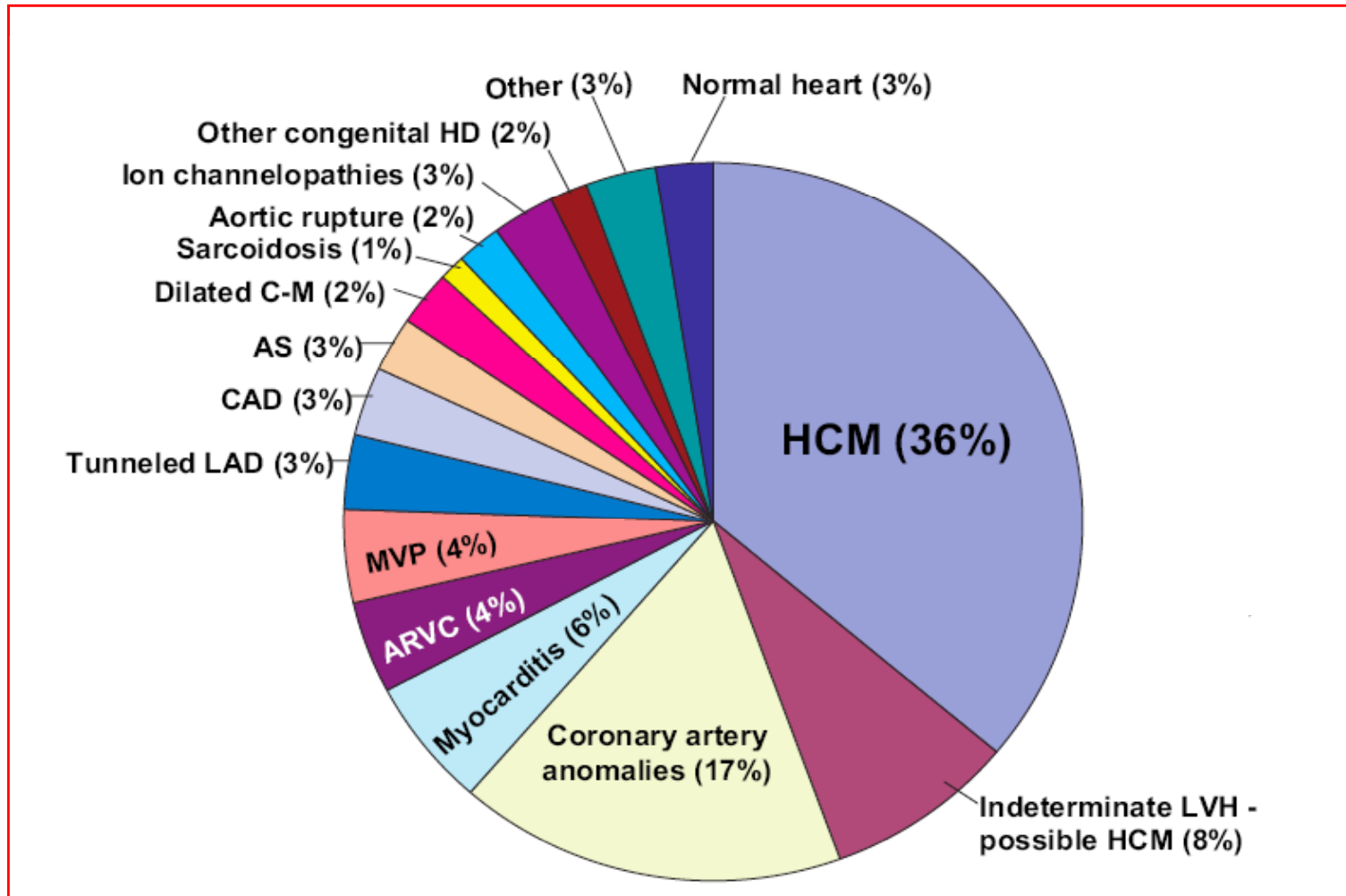


# Causes of SCD

- Overall > 80 % Coronary heart disease
- 10-15% Non-ischemic structural heart disease
  - Cardiomyopathies
  - Myocarditis
  - Congenital heart disease
  - Aortic rupture
- 5% Structurally normal heart (SADS)



# International variation : Causes of SCD in US athletes



Maron et al. Circulation March 2007 (online)



## Causes of SCD in Italy 1979-1998

273 consecutive cases in Veneto region evaluated

	<b>Number</b>	<b>%</b>
Abnormal hearts	197	72
Obstructive coronary disease	54	27
Valve disease	31	16
Coronary anomalies	28	14
ARVC	27	13
HCM	18	9
Aortic rupture	13	7
DCM	12	6
Other	14	7
Apparently normal hearts	76	
- Concealed pathology	60	28
- ARVC	9	30 (79% of 'normals')
- Conduction system disease	24	

Adapted from *D. Corrado et al. / Cardiovascular Research 50 (2001) 399 –408*



## Hereditary diseases detectable on screening

- Hypertrophic Cardiomyopathy (HCM)
- Dilated Cardiomyopathy (DCM)
- Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC or ARVD)
- Marfan's disease
  
- Long QT
- Brugada syndrome
- Short QT



# Epidemiology

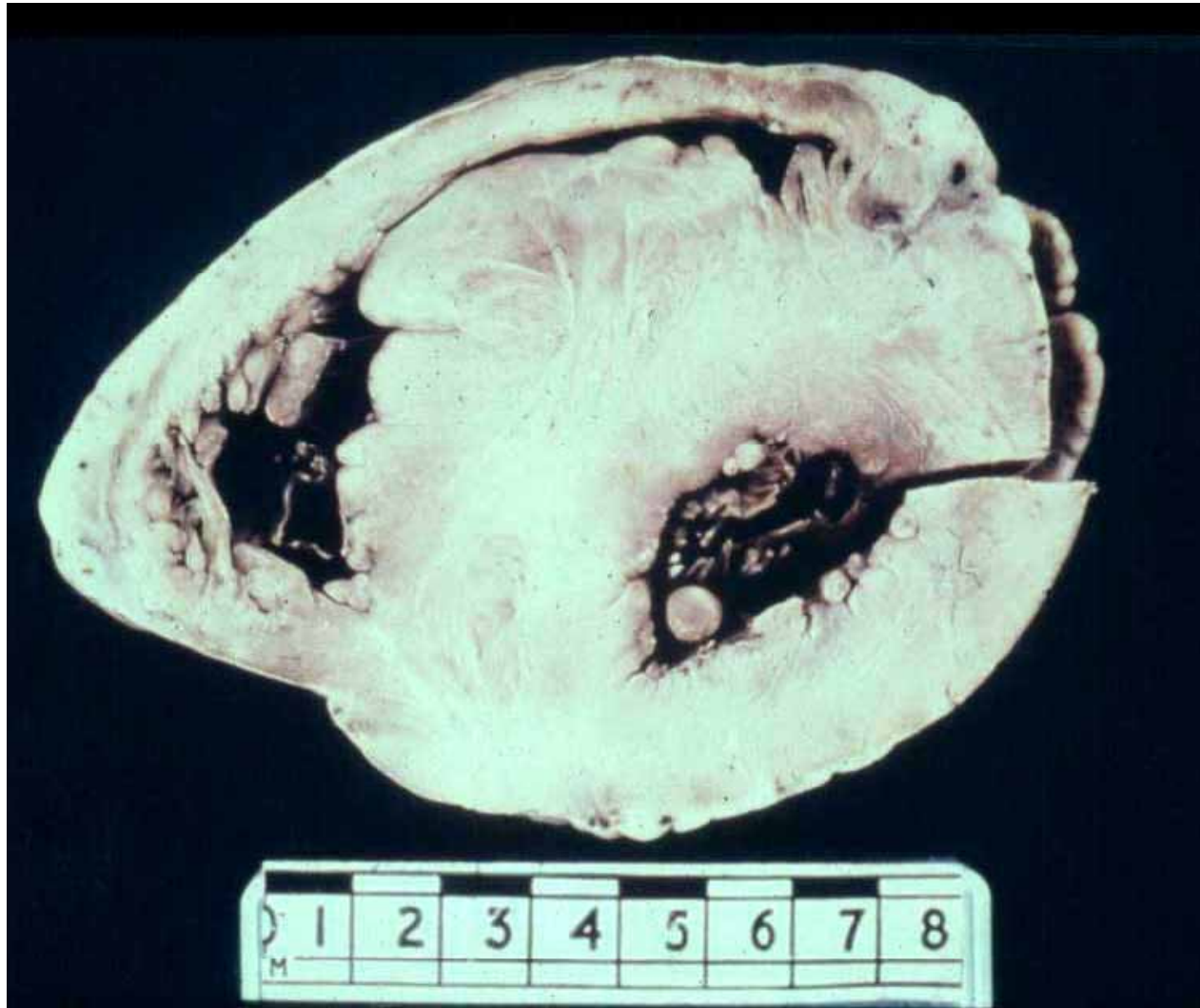
## Prevalence

## Estimated number affected in Ireland

HCM	1:500	9,000
ARVC	1:1,000 - 10,000	900
DCM	1:3,000 - 5,000	900-1500



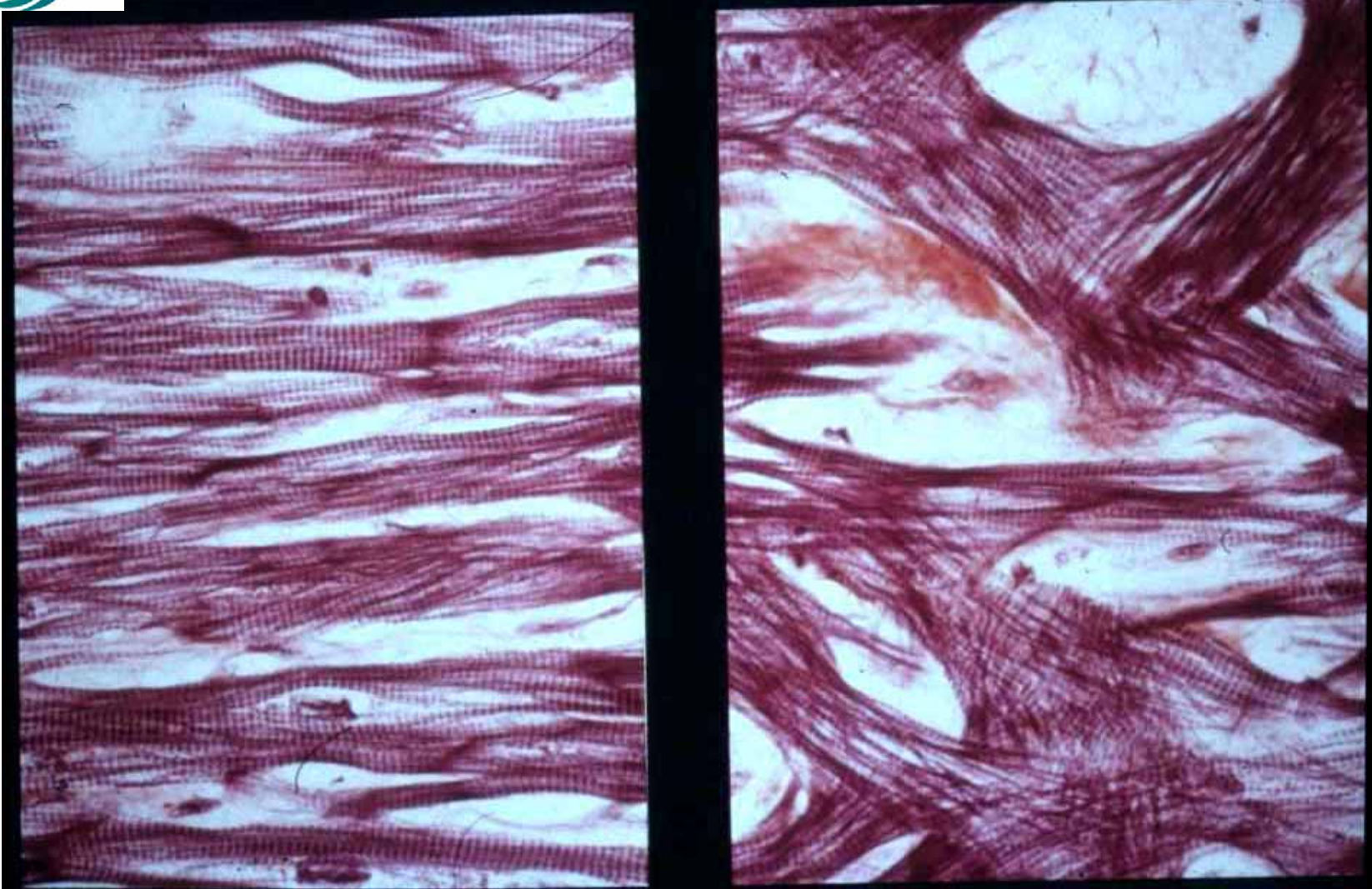
# Hypertrophic Cardiomyopathy



**Dx – unexplained LVH**



# Hypertrophic Cardiomyopathy



Histology - normal

Dx – HCM (hypertrophy + disarray)

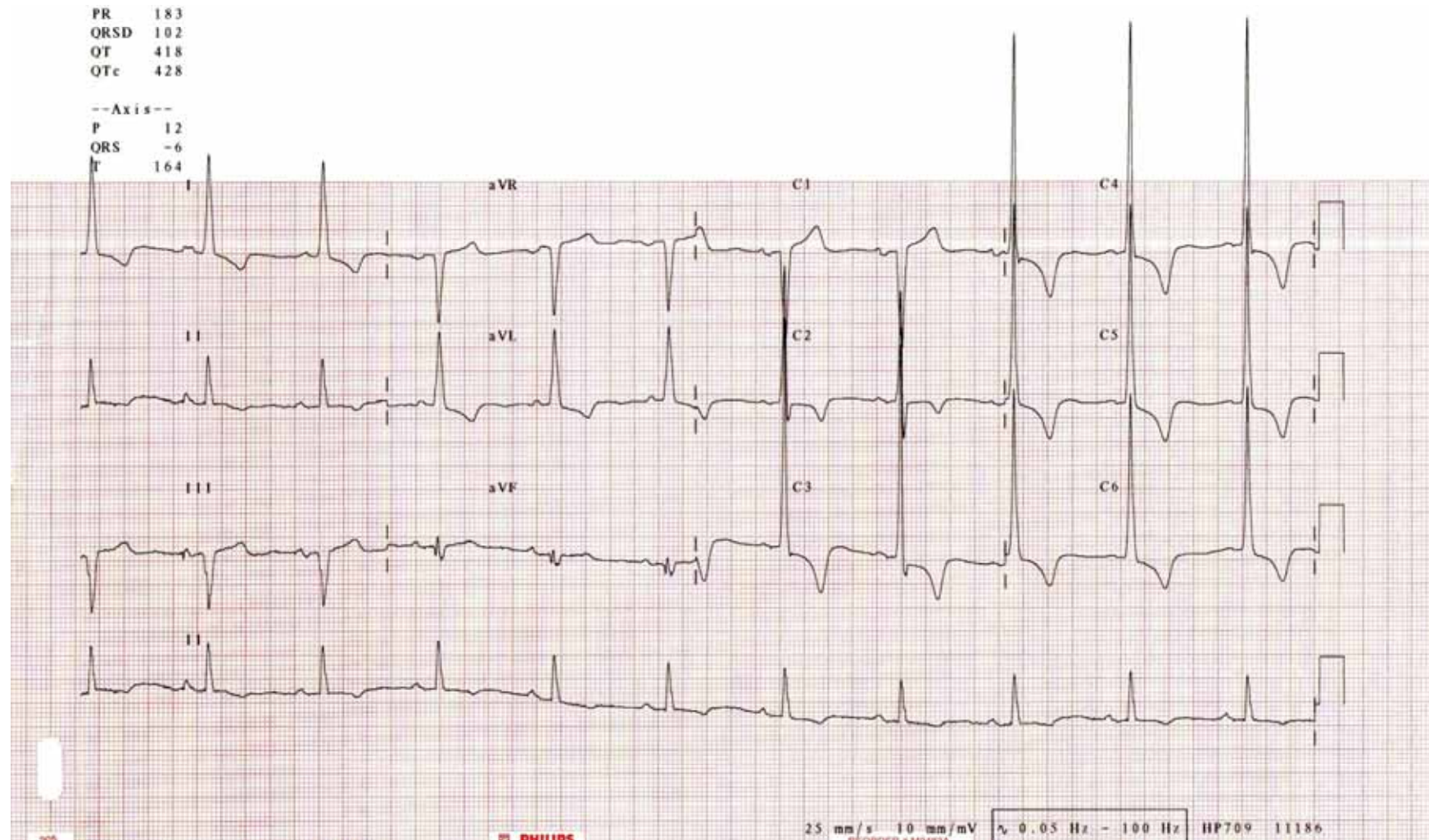


# PRESENTATION OF HCM

- Symptoms :
  - Chest pain
  - Shortness of breath
  - Palpitations
  - Pre-syncope / syncope
  - Cardiac arrest
- No symptoms :
  - Incidental finding
  - Family screening

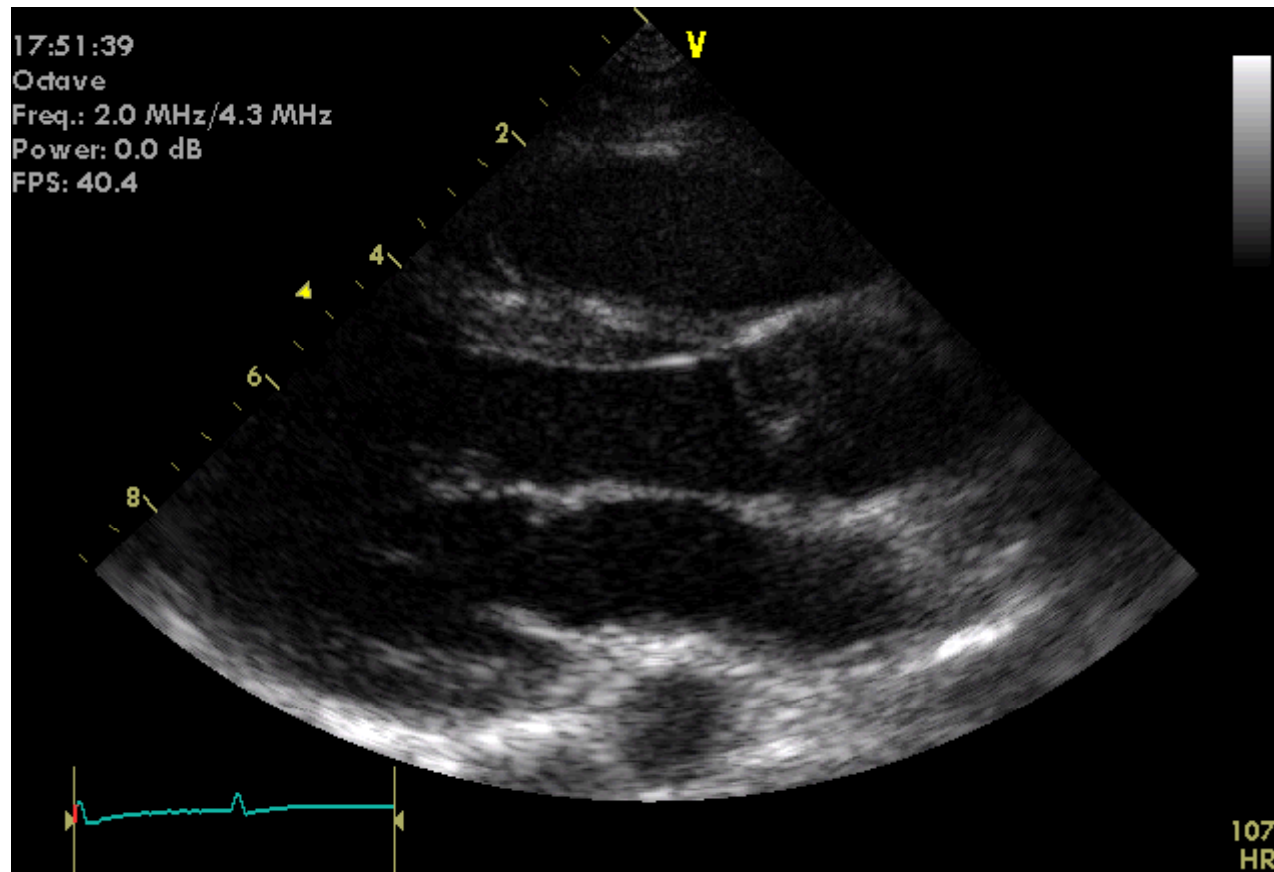


# HCM ECG



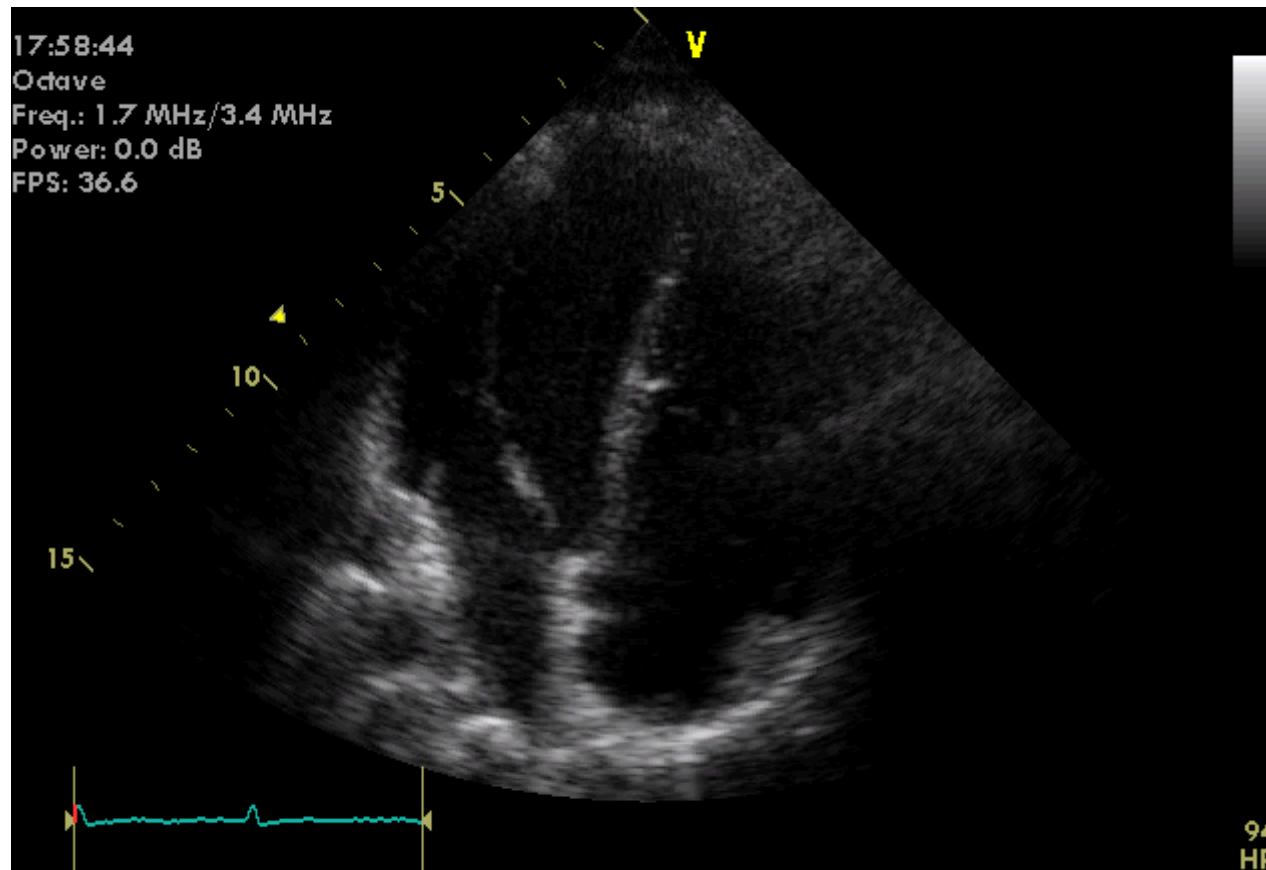


# Normal Echo



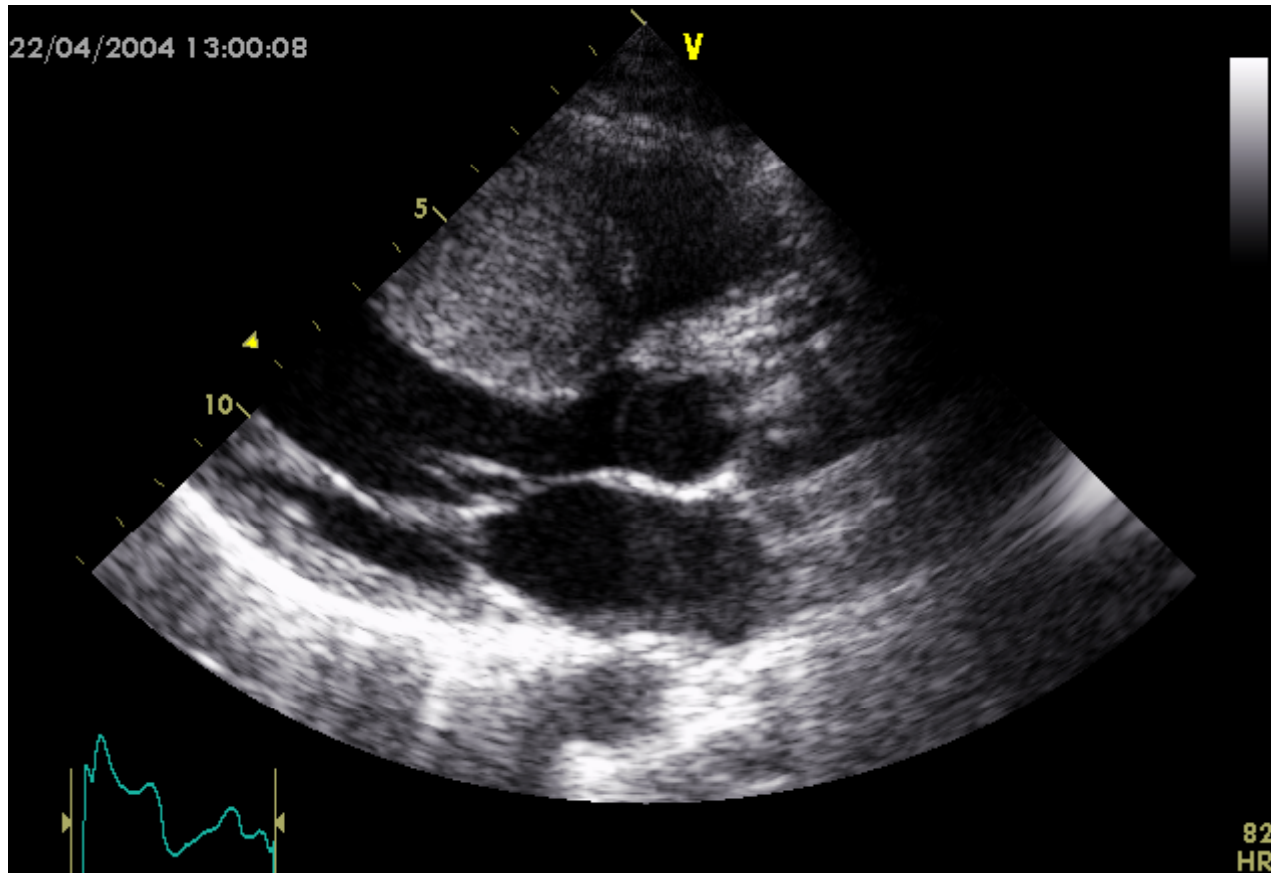


# Normal echo (apical view)



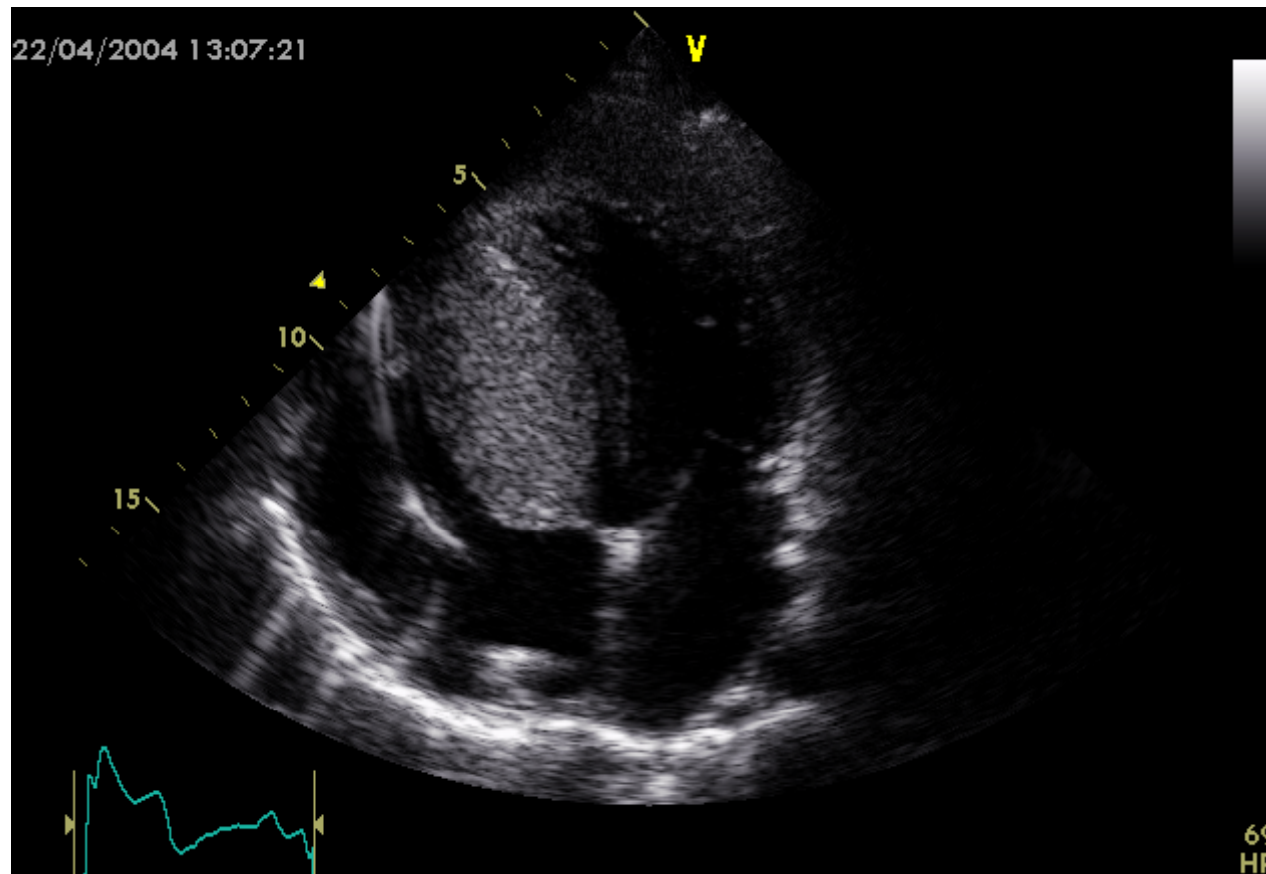


# HCM Echo





# HCM Echo (apical view)





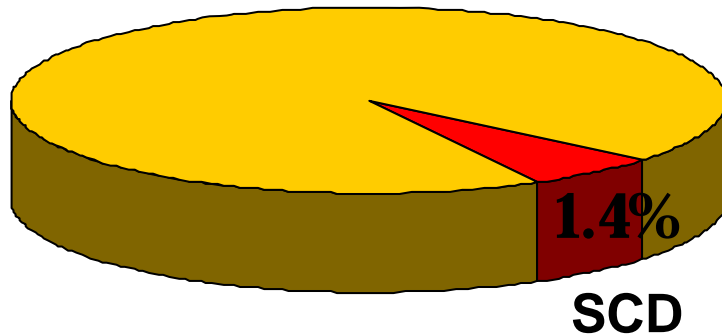
# Complications of HCM

- None
- Chest pain (? Microvascular disease)
- LVOT obstruction
- Atrial fibrillation
- Thrombo-embolic events
- Heart failure
- Cardiac arrest

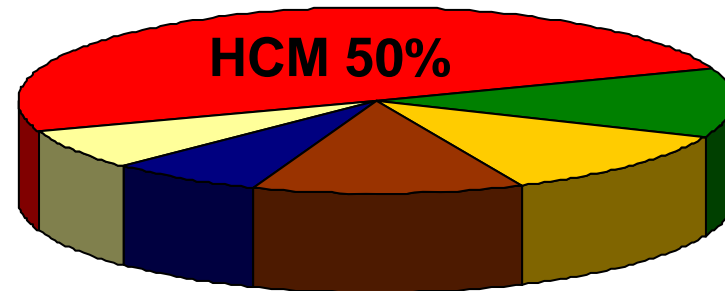


# Sudden Cardiac Death in HCM

## Magnitude of the Problem



*Percentage SCD  
in HCM*



*Causes of SCD  
in the Young*

**N = 5413, f/u = 6 yrs**  
**SCD = 1.4% / yr**



# Clinical risk factors

- Features that may indicate increased risk :
  - Previous cardiac arrest
  - Family history premature sudden death
  - Unexplained blackouts
  - VT or NSVT on exercise test or Holter monitor Blood pressure fails to rise normally with exercise
  - Severe thickening of the heart (>3 cms or almost 3x normal)
- Some more significant at younger age
- Presence of LVOTO may also increase risk
- No risk factors < 1% death per year
- 2 risk factors = 3% risk, 3 or > = 6% + risk



# Dilated Cardiomyopathy (DCM)

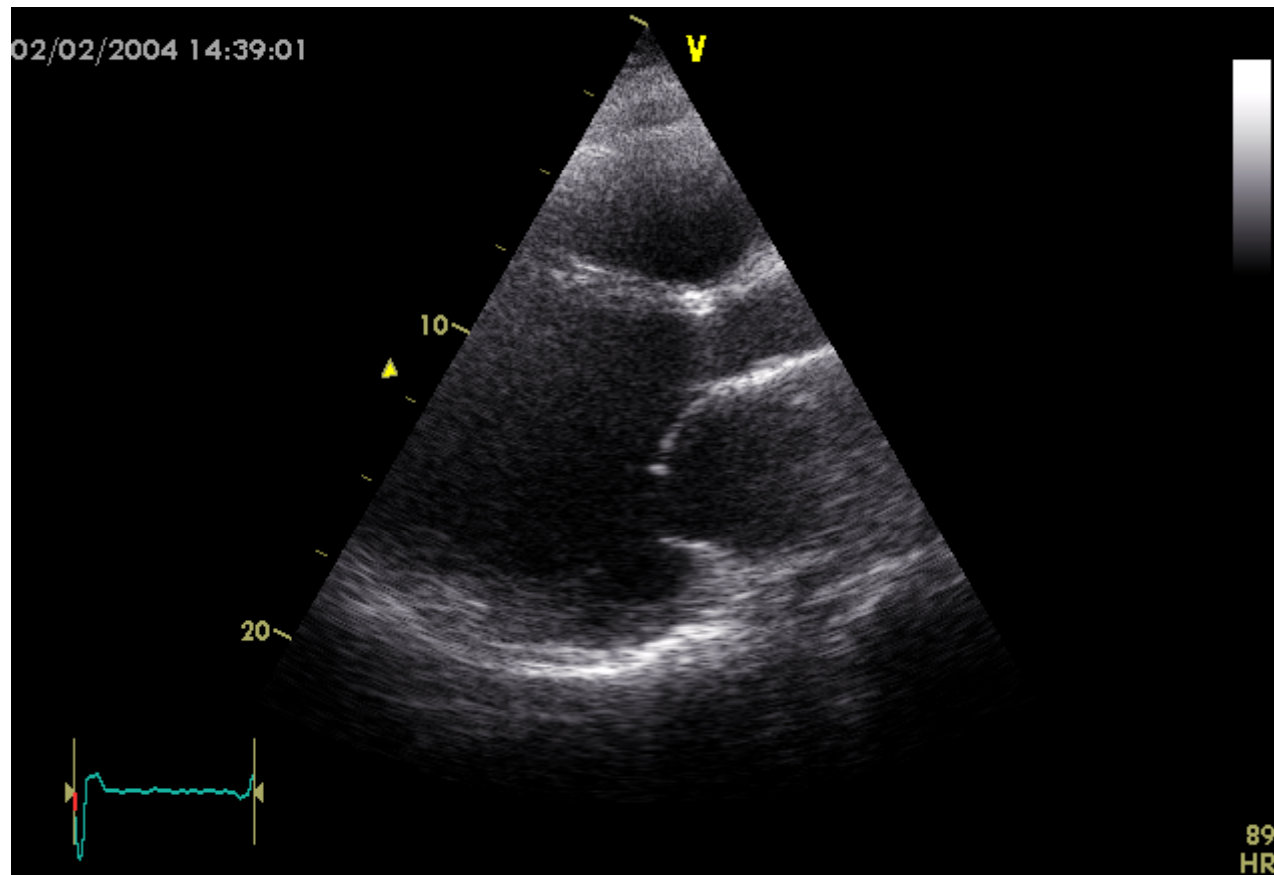
## – Causes of heart failure

- IHD
- Valvular heart disease
- Hypertension
- Viral
- Alcohol
- Infectious
- Drugs
- 'High-output' (TFTs, anaemia, incessant tachy)
- Idiopathic





# DCM echo





# DCM Presentation

- Symptomatic :
  - Shortness of Breath
  - Fatigue
  - Reduced Exercise tolerance
  - Palpitations
  - Pulmonary oedema
- Asymptomatic :
  - Unmasked during procedure
  - Executive / Insurance Health Screen
  - Family Screen



# Diagnosis of DCM

- History (recent virus, symptoms, drug usage, family history)
- ECG
- ECHO
- Coronary Angio
- + / - :
  - Viral screen
  - Skeletal muscle biopsy / metabolic screen
  - RV biopsy
  - MRI



## Arrhythmogenic right ventricular cardiomyopathy (ARVC / ARCD)

- Commonest cause of SCD in athletes in Northern Italy.
- Prevalence 1:1000 (?)
- Familial in 30-40% of cases.
- Fibrofatty replacement of the right ventricle.
- Fatal ventricular arrhythmias of right ventricular origin.
- 5 genes identified (account for < 50%)

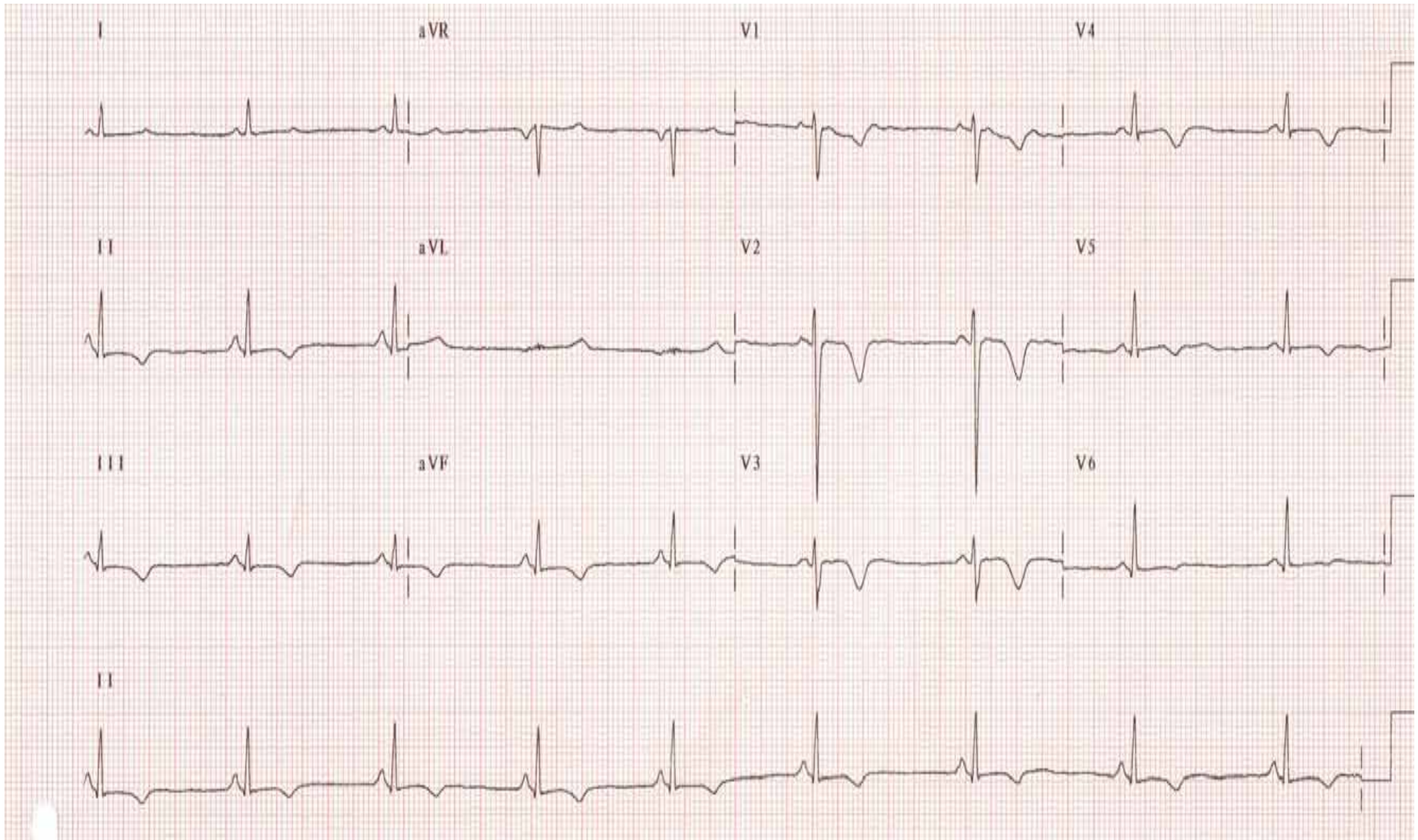


# DIAGNOSIS ARVC

- ECG
- SAECG
- Transthoracic Echo (conventional)
- Holter monitoring
- Exercise testing
- MRI
- 'Special' Echo (Tissue Doppler Imaging, Contrast Echo, 3-D Echo)
- Genetic testing



# Typical ARVC ECG





# Sudden Arrhythmic Death Syndrome (SADS)

- Cause not apparent on PM
- Cave potentially spurious causes
  - Non-obstructive coronary disease with no infarct
  - ‘LVH’ with normal heart weight
  - ‘sudden death in epilepsy’
- 40% of families have inherited cause identified (mostly LQT and Brugada)



# Long QT syndrome

- Prevalence may be 1 in 5000
- Autosomal dominant
- Not detectable after death (unless DNA)
- Abnormal T-wave morphology +/- QT prolongation
- Genetic mutation detected in 80%
- Treatment
  - Beta-blockers (not in LQT2)
  - ICD

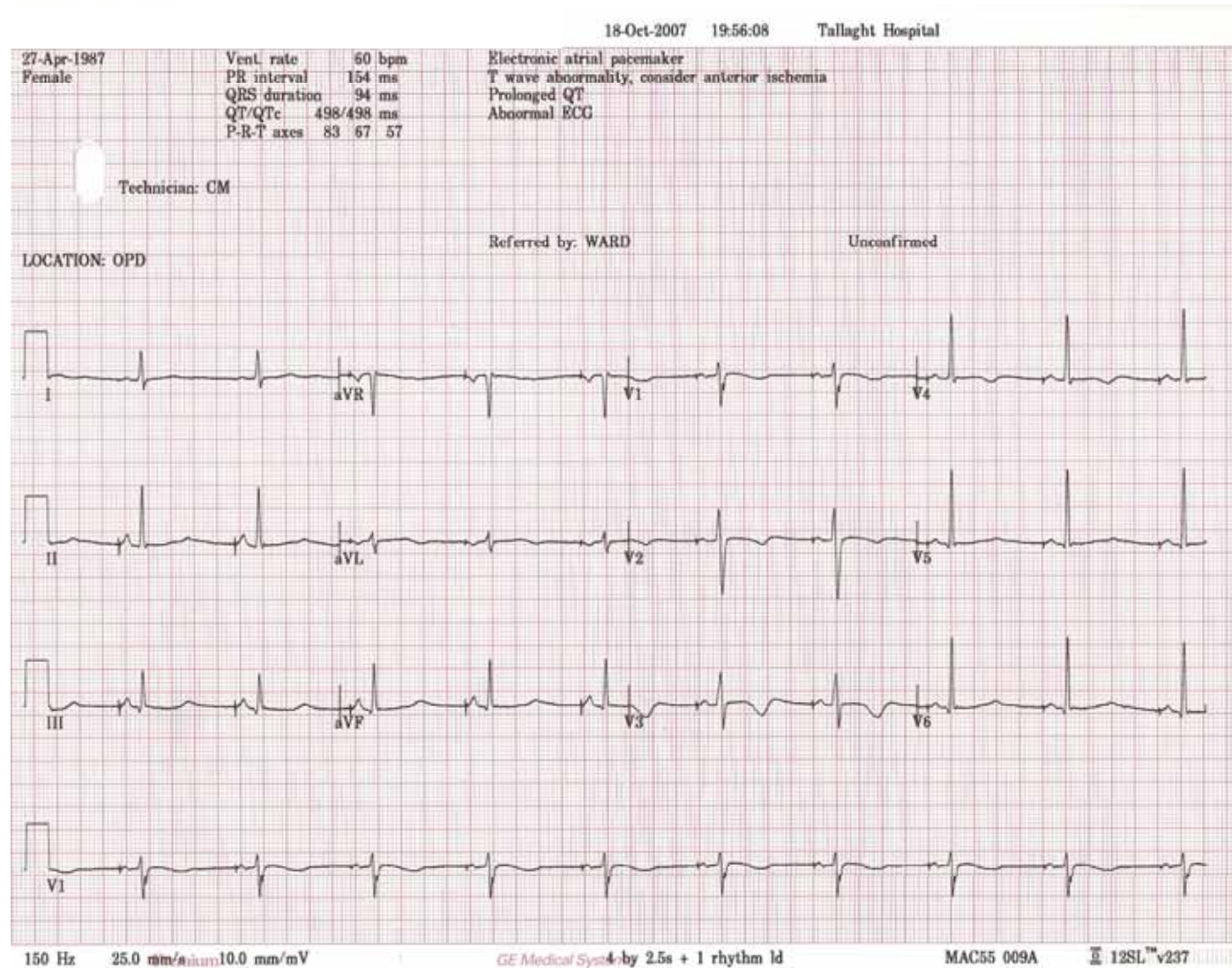


# LQTS

- QTc tends to be higher from LQT1 to LQT3
- QTc was higher in patients who experienced events across all 3 groups
- % gene positive patients with normal QTc
  - 36% LQT1
  - 19% LQT2
  - 10% LQT3



# Typical ECG



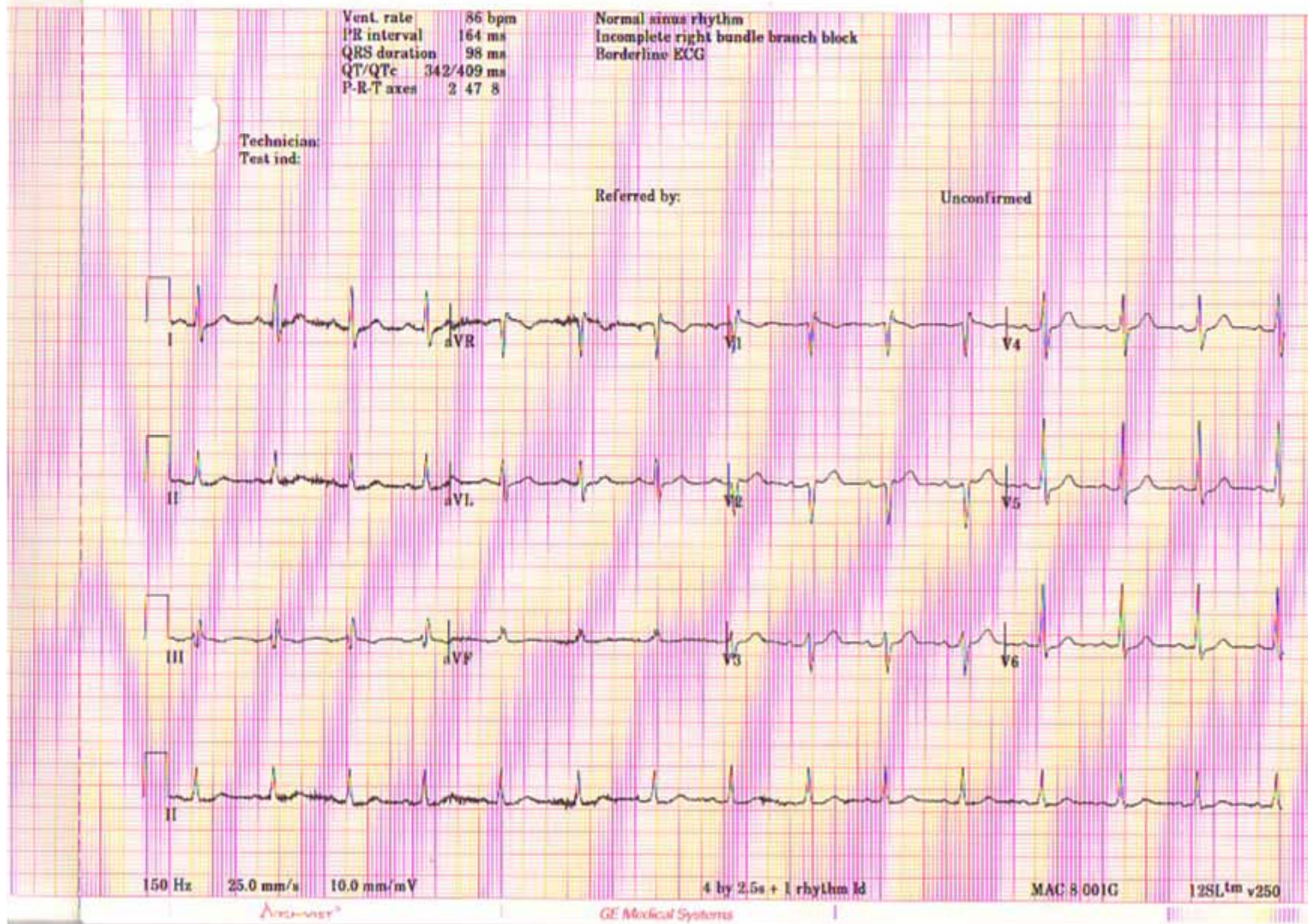


# Brugada syndrome

- True prevalence unknown
- Ventricular arrhythmia (polymorphic VT) + ST elevation V1-V3
- Sodium channel mutation found in 20%
- 4 other genes recently discovered
- Characteristic ECG not always present at rest
  - May be unmasked by infusion of Na-channel inhibitors (Flecainide / Ajmalin)
- Management
  - VT stim
  - ICD

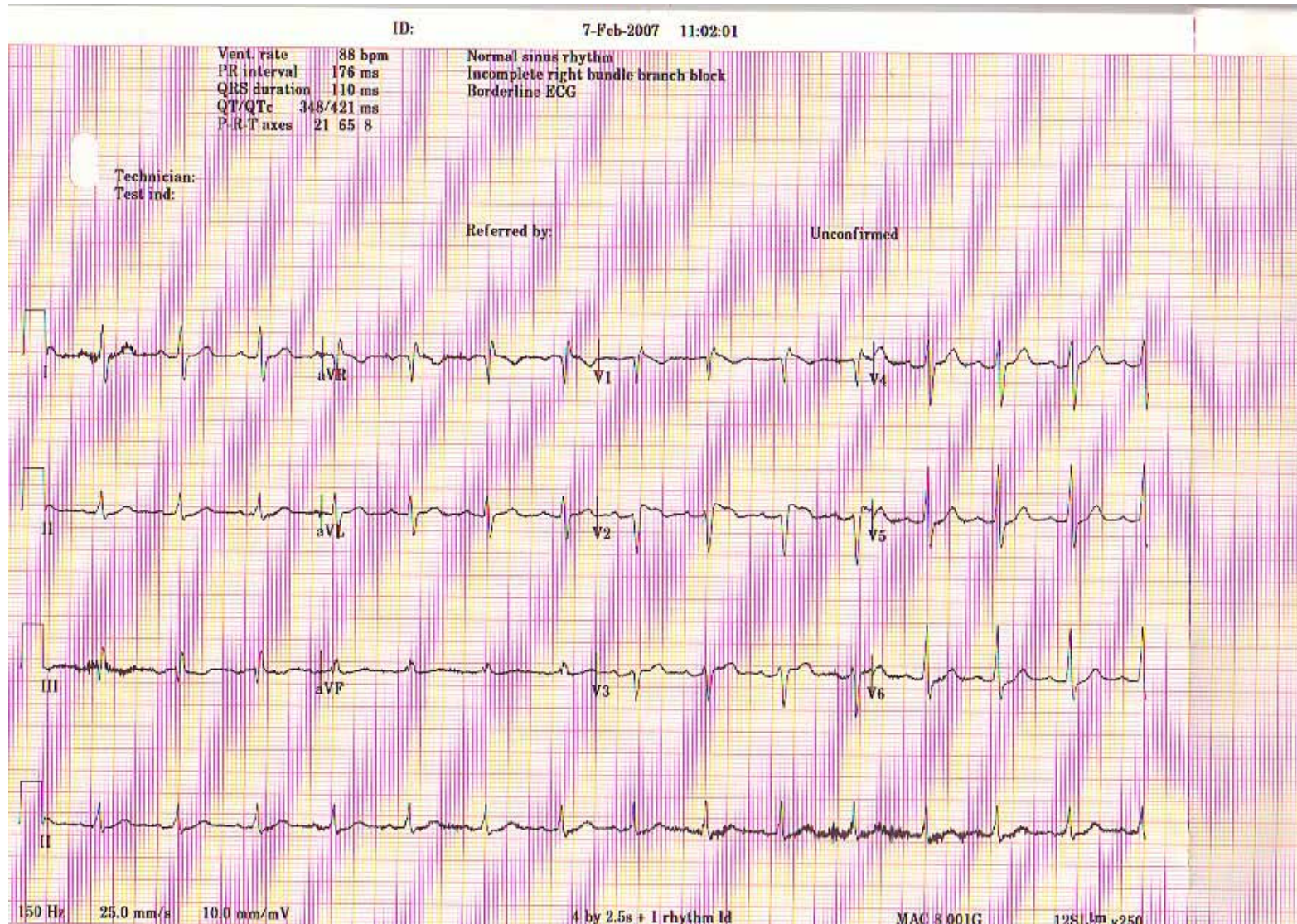


# Patient pre - Ajmalin





# Patient during Ajmalin





# In summary

- SCD common in older age groups
- Majority of cases secondary to IHD
- Younger age groups inherited cardiac disease more prominent
- Careful post-mortem required in all SCD cases
- Family evaluation advised in all cases where definite or possible inherited cardiac disease