APCDE 2011

Non-compaction & Other Unusual Cardiomyopathies

Dae-Hee Kim, MD, PhD
College of Medicine
University of Ulsan
Asan Medical Center

ASAN Medical Center

UNIVERSITY OF ULSAN COLLEGE OF MEDICINE

Historical context

- **1957 : the term " Cardiomyopathy" for the first time ; primary myocardial disease**

- **1968 (WHO): "Diseases of different and often unknown etiology in which the dominant feature is cardiomegaly and heart failure"**

- **1980 (WHO) : "Heart muscle diseases of unknown cause"**

Classification (1)

- 1995 WHO/ISFC definition & classification
  - Diseases of the myocardium associated with cardiac dysfunction
  - Dilated cardiomyopathy
  - Hypertrophic cardiomyopathy
  - Restrictive cardiomyopathy
  - Arrhythmogenic right ventricular cardiomyopathy
  - Unclassified cardiomyopathies

Classification (2)

Cardiomyopathies

HCM

Familial
- Sacomere protein mutation
- Storage disease
- Disorders of fatty acid metabolism
- Carnitine deficiency
- Phosphorylase B kinase deficiency
- Mitochondrial cytopathies
- Syndromic HCM
- Ohters

Non-familial
- Obesity
- Infants of diabetic mothers
- Athletic training
- Amyloid (AL/prealbumin)

DCM

Familial
- Sacomere protein mutation
- Z-band
- Cytoskeletal genes
- Nuclear membrane
- Mildly dilated CM
- Intercalated disc protein mutations (see ARVC)
- Mitochondrial cytopathy

Non-familial
- Myocarditis
- Kawasaki disease
- Eosinophilic (Churg Strauss syndrome)
- Viral persistence
- Drugs
- Pregnancy
- Endocrine
- Nutritional
- Alcohol
- Tachycardio myopathy

ARVC

Familial
- Intercalated disc protein mutations
- Cardiac ryanodine receptor (RyR2)
- Transforming growth factor-β3 (TGFβ3)

Non-familial
- Inflammation

RCM

Familial
- Sarcomeric protein mutations
- Familial amyloidosis
- Desminopathy
- Pseuxanthoma elasticum
- Haemochromatosis
- Anderson-Fabry disease
- Glycogen storage disease

Non-familial
- Amyloid (AL/prealbumin)
- Scleroderma
- Endomyocardial fibrosis
- Carcinoid heart disease
- Metastatic cancers
- Radiation
- Drugs (anthracyclines)

Unclassified

Familial
- Left ventricular non-compaction
  - Barth syndrome
  - Lamin A/C
  - ZASP
  - α-dystrobrevin

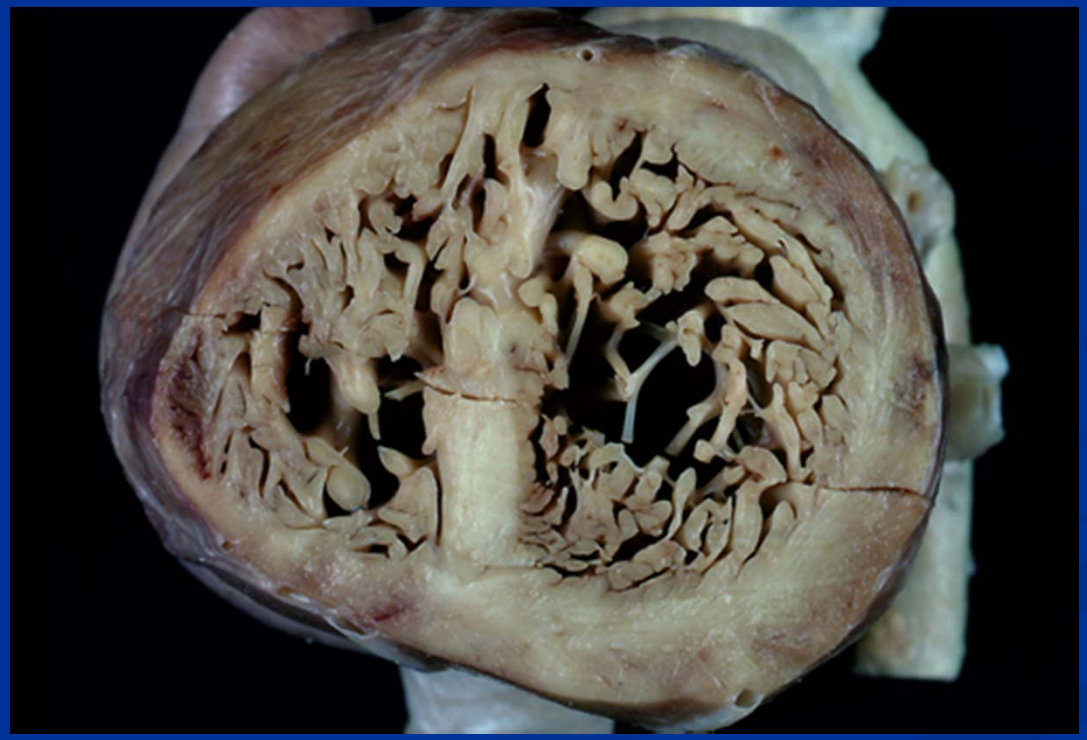
Non-familial
- Tako Tsubo cardiomyopathy

2008 ESC recommendation

Agenda

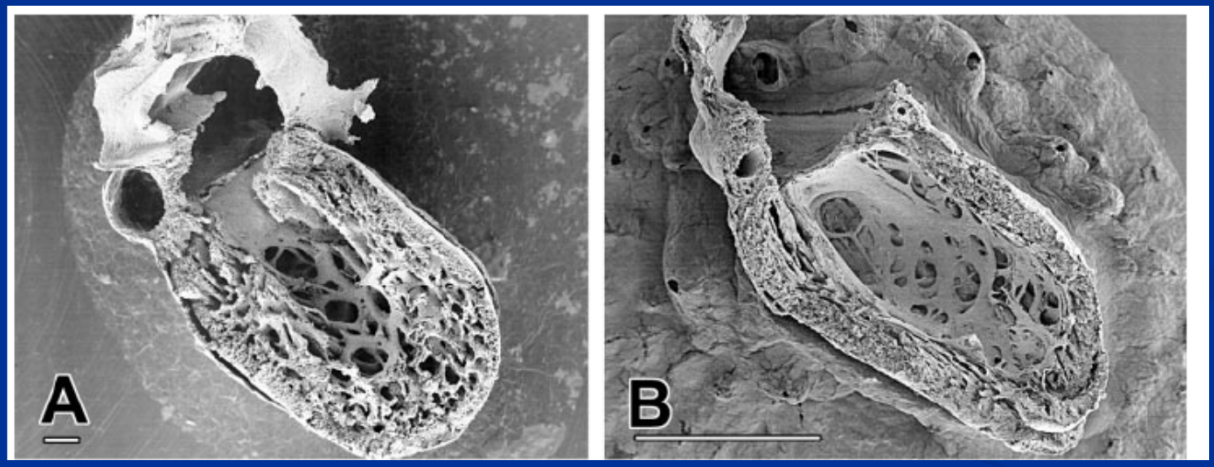
- LV non-compaction cardiomyopathy
- Arrhythmogenic RV cardiomyopathy
- Stress-induced cardiomyopathy

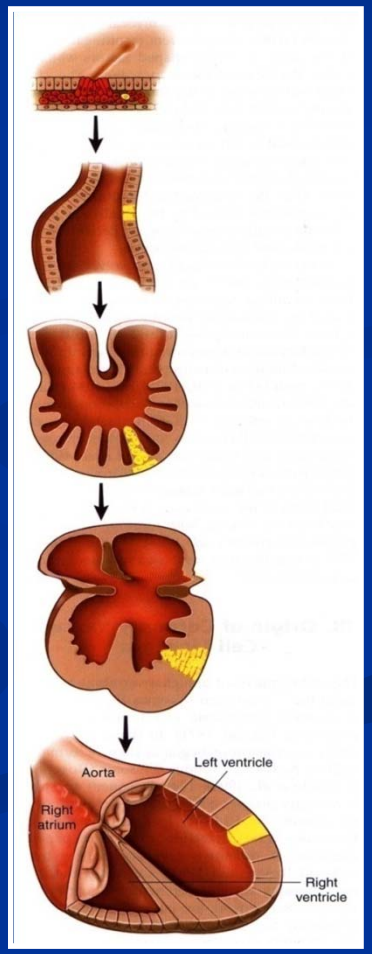
Non-Compaction Cardiomyopathy



Noncompaction cardiomyopathy

- Compaction of the ventricular myocardium normally progresses from epicardium to endocardium and from the base of the heart toward the apex : 5-8 weeks of fetal development

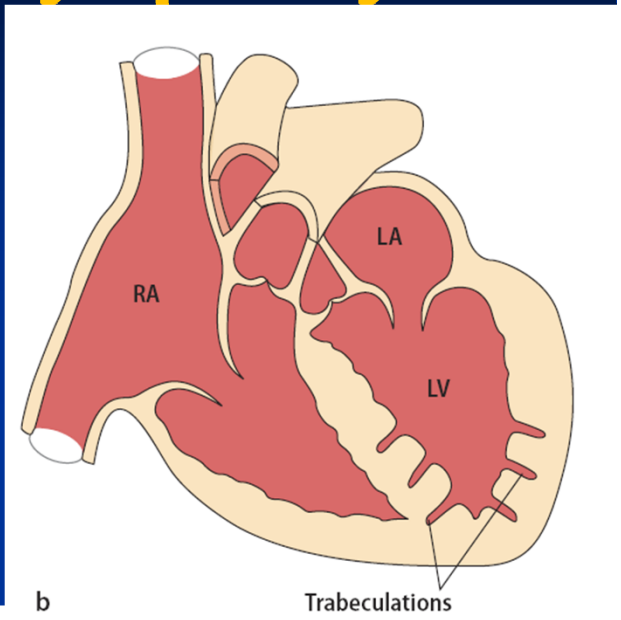




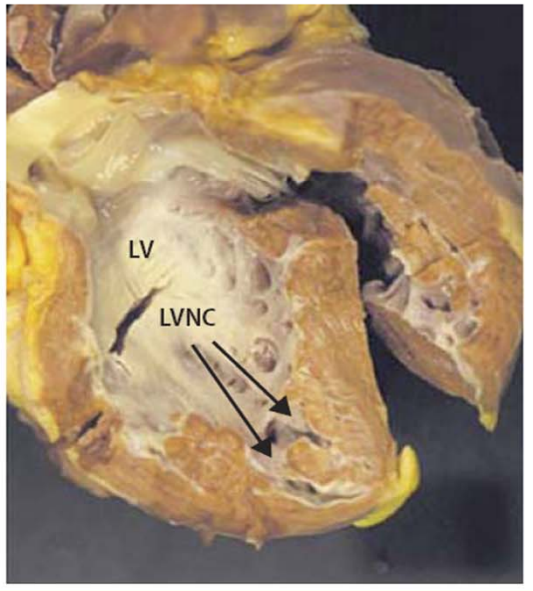
- Arrest of compaction, non-compaction of normal myocardium: result in persistence of the deep recesses and sinusoids

Noncompaction cardiomyopathy

- Segments of noncompacted myocardium mainly involve the apex and the inferior mid and lateral mid of the left ventricular wall

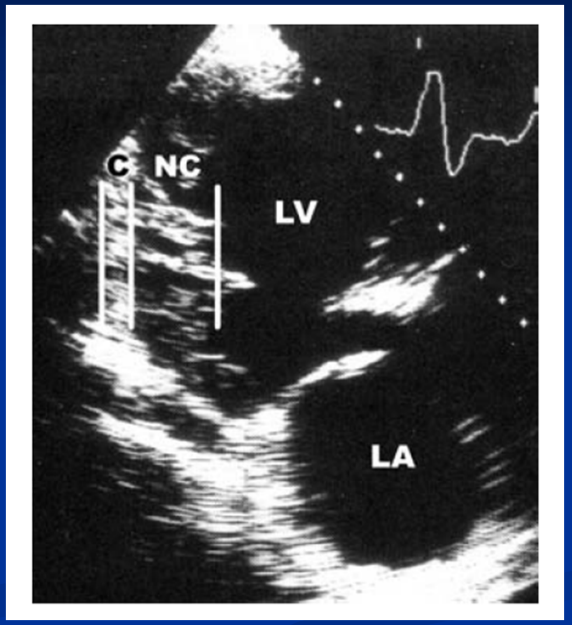


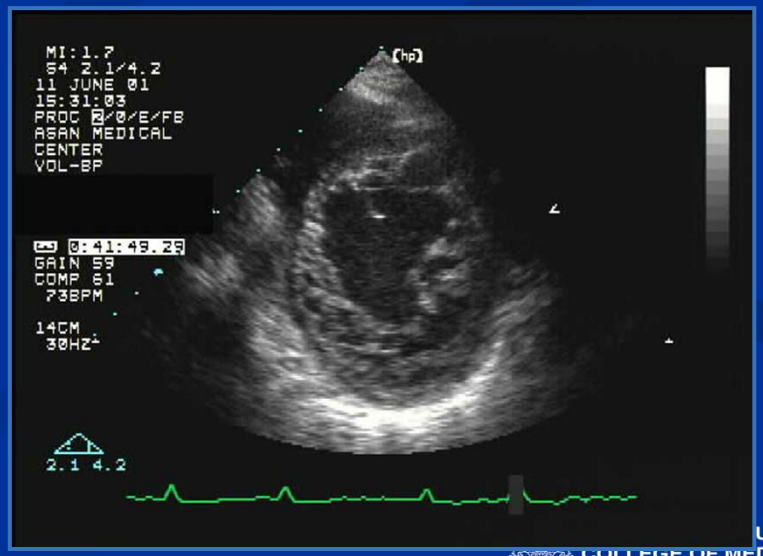
	Pediatric patients	Adult patients	
	Ichida [21]	Oechslin [31]	Sengupta [36]
LV apex	100%	94%	100%
LV inferior wall	70%	94%	95%
LV lateral wall	41%	100%	100%
Basal segments of the LV wall		< 20%	< 27%

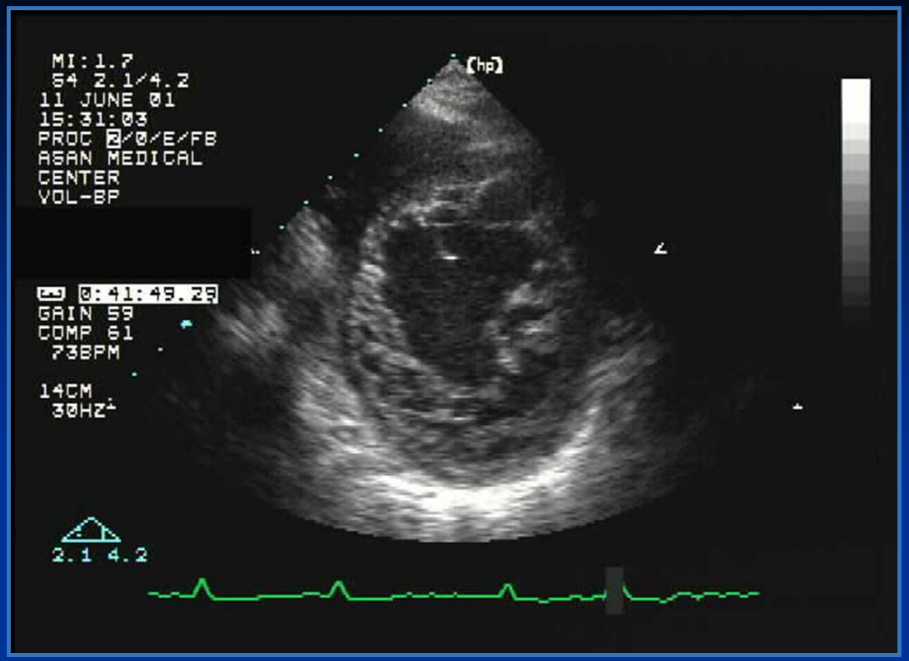


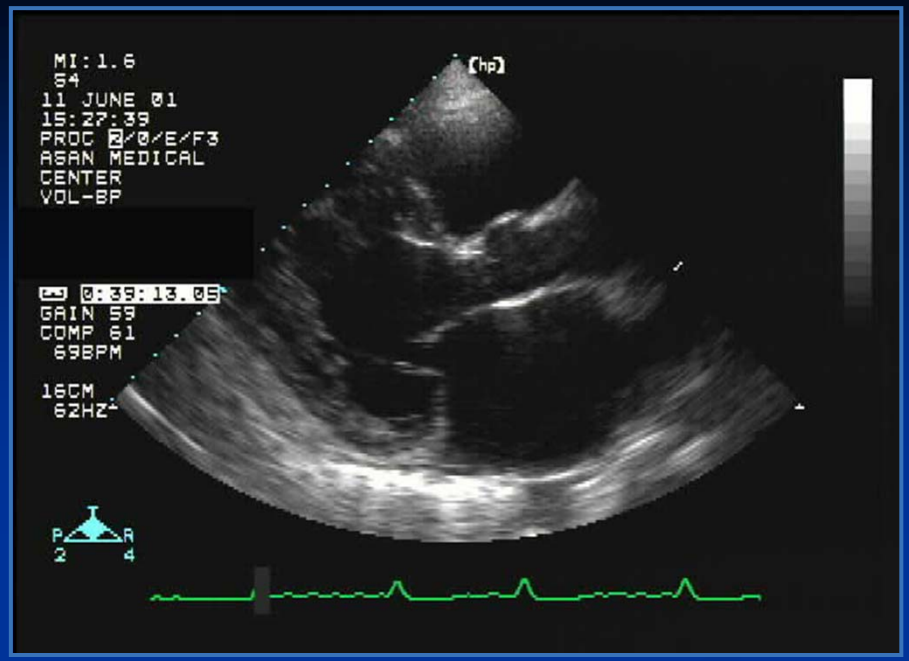
Diagnostic Criteria of LV Non-compaction

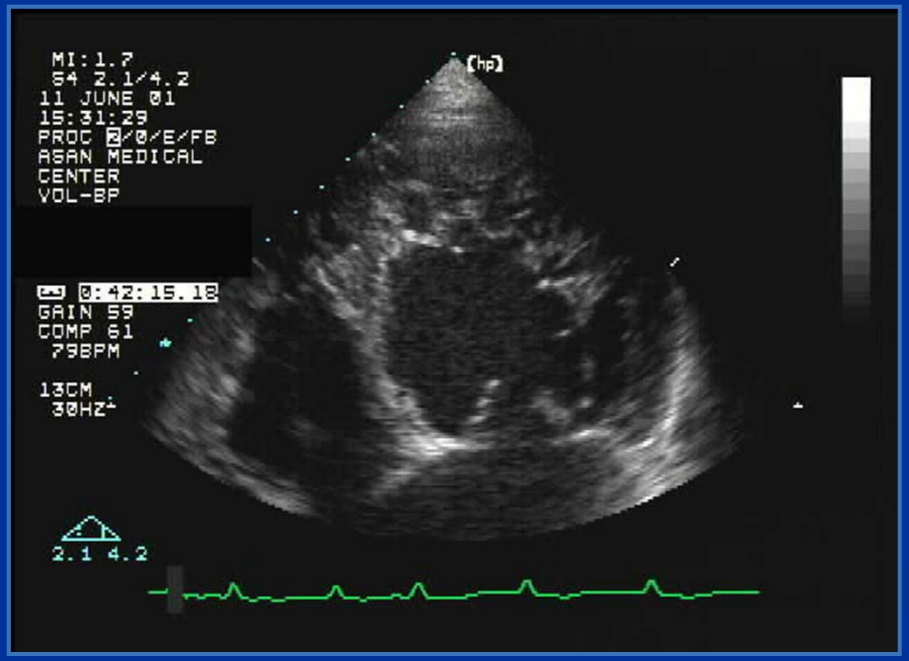
- Appearance of at least four prominent trabeculations and deep intertrabecular recesses
- Endsystolic noncompaction/compaction ratio (maximal thickness site) ≥ 2
- Deep recesses filled with blood from the ventricular cavity visualized by color Doppler imaging

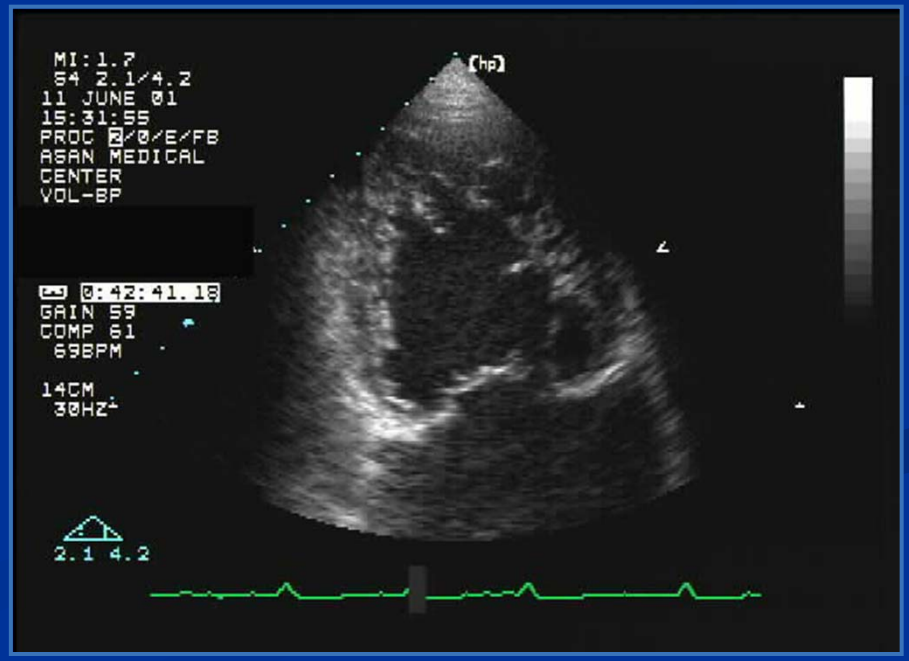




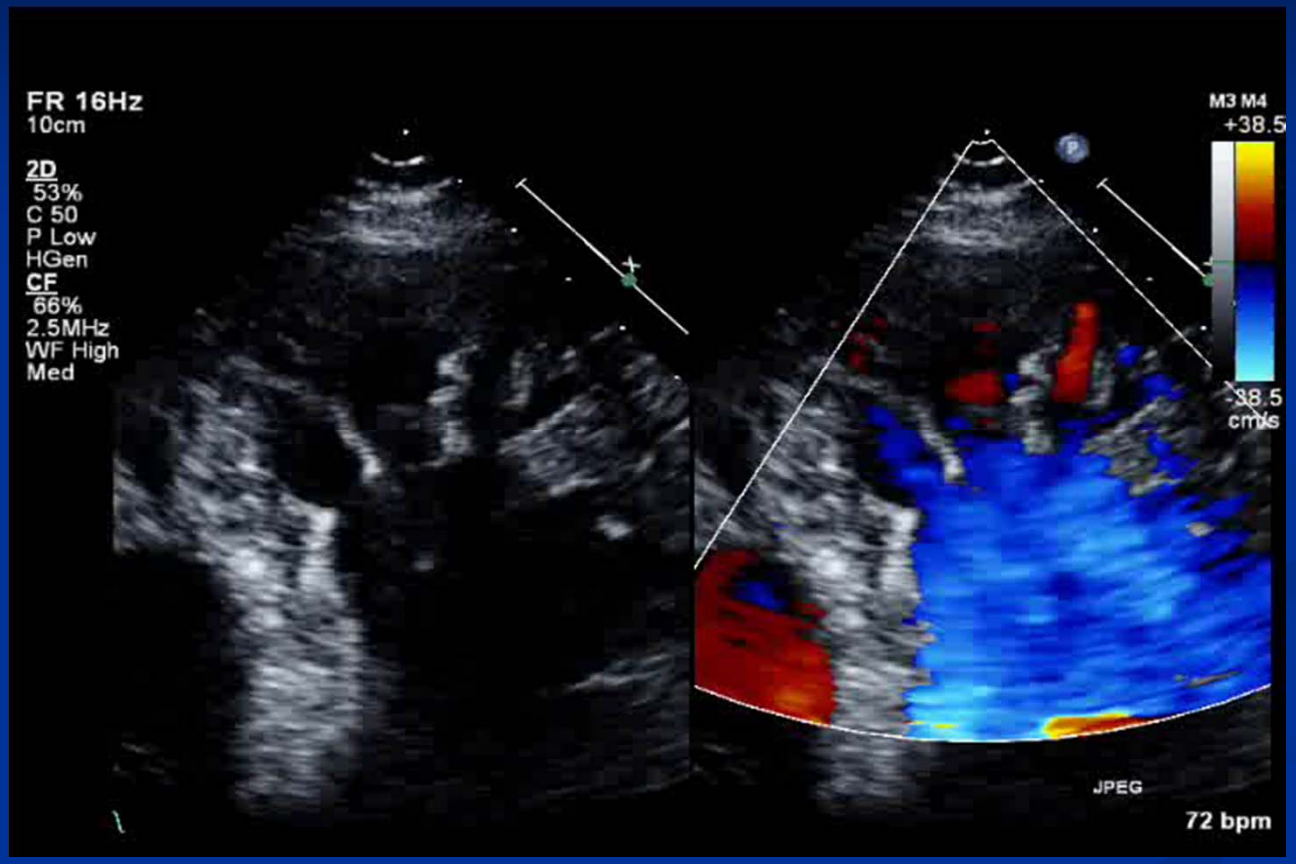

MI:1.7
S4 2.1/4.2
11 JUNE 01
15:31:03
PROC 2/0/E/FB
ASAN MEDICAL
CENTER
VOL-BP
0:41:49.29
GAIN 59
COMP 61
73BPM
14CM
30HZ
2.1 4.2


MI:1.6
S4
11 JUNE 01
15:27:39
PROC 2/0/E/F3
ASAN MEDICAL
CENTER
VOL-BP
0:35:13.05
GAIN 59
COMP 61
69BPM
16CM
62HZ
P R
2 4


MI:1.7
S4 2.1/4.2
11 JUNE 01
15:31:29
PROC 2/0/E/FB
ASAN MEDICAL
CENTER
VOL-BP
0:42:15.18
GAIN 59
COMP 61
79BPM
13CM
30HZ
2.1 4.2


MI:1.7
S4 2.1/4.2
11 JUNE 01
15:31:55
PROC 2/0/E/FB
ASAN MEDICAL
CENTER
VOL-BP
0:42:41.18
GAIN 59
COMP 61
69BPM
14CM
30HZ
2.1 4.2

Fireworks in the Left Ventricle: Doppler Manifestation of Left Ventricular Noncompaction



- Deep recesses filled with blood from the ventricular cavity visualized by color Doppler imaging

Kim et al. JACC 2009

Prevalence of Left Ventricular Noncompaction

- True prevalence : unclear, selection bias
- A review from Switzerland identified 34 cases in 15 years
- Prevalence: 0.014% of patients referred to Echo lab

Oechslin et al. JACC 2000

- Male predominance
- 3.7% prevalence for LVEF <45%

Sandhu et al. Echocardiography 2007

- Association with facial dysmorphism & NMD

Left Ventricular Noncompaction

- LVNC is associated with Neuromuscular diseases : as many as 82% of patients
- 62 patients with LVNC underwent neurological evaluation
  - Metabolic myopathy
  - Myotonic, Becker, Duchenne : (43%)
  - NMD of unknown etiology (39%)
  - Neurologically normal (18%)

Stollberger et al. AJC 2002

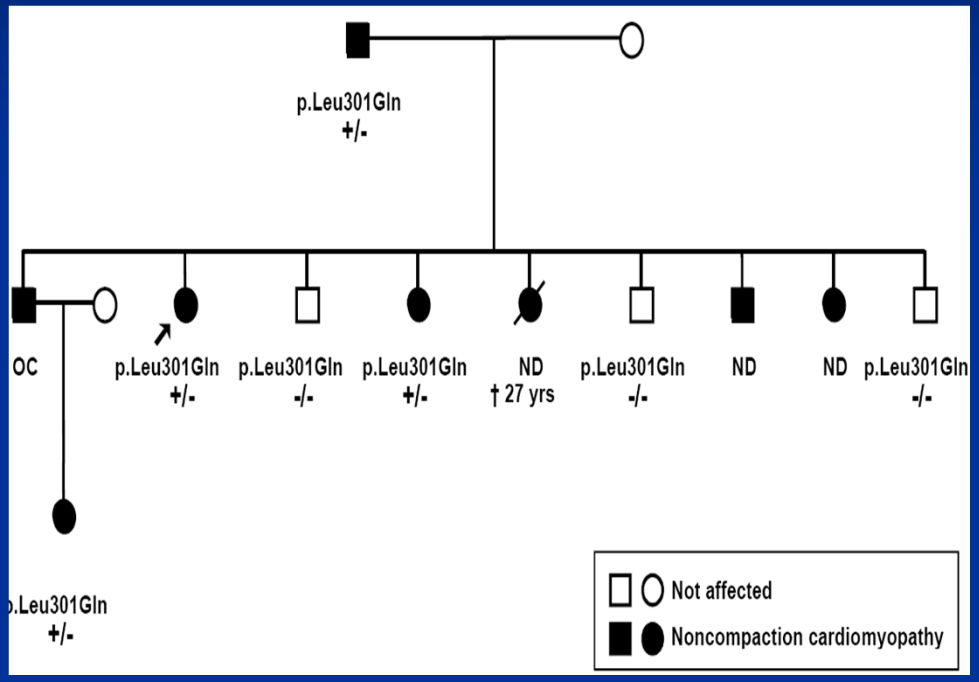
Clinical presentation

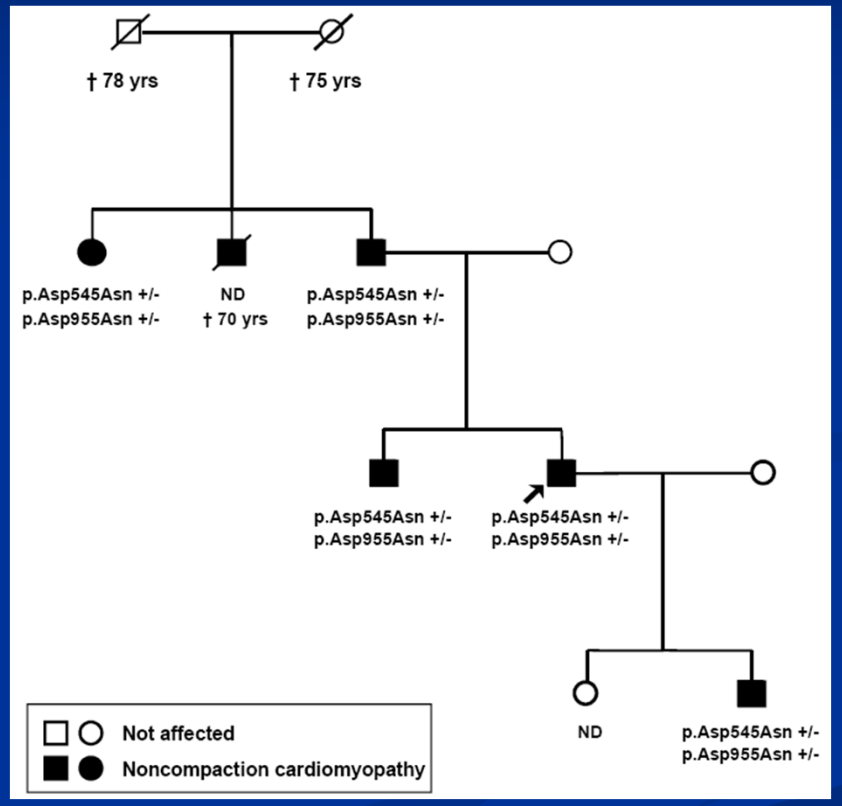
- Heart failure
- Thromboembolic event
- Arrythmias : VT, AF

Characteristics	Pediatric patients		Adult patients			
	Chin [9]	Ichida [21]	Oechslin [31]	Sengupta [36]	Murphy [30]	Stollberger [39]
Number of patients	8	27	34	32	45	62
Age at diagnosis (median)	7 yrs	5 yrs	40 yrs	49 yrs	37 yrs (mean)	50 yrs (mean)
Male	63%	56%	74%	53%	62%	79%
Familial occurrence	50%	44%	18%			
Follow-up	up to 5 yrs	up to 17 yrs	up to 11 yrs		up to 15 yrs	up to 6 yrs
ECG						
Bundle branch block	25%	15%	56%		29%	23%
WPW-syndrome	13%	15%	0%			3%
Ventricular tachycardia	38%	0%	41%		20% a	18%
Clinical symptoms						
Heart failure symptoms	63%	30%	68%	62.50%	62%	73%
Systemic embolic events	38%	0%	21%		4%	
Pulmonary embolism	0%	7%	9%			
Ventricular thrombi	25%	0%	9%	6%		
Facial dysmorphism	38%	33%	0%			
Neuromuscular disorders						82%
Deaths	38%	7%	35%		2%	
Heart transplantation	0%	4%	12%			

MYH7 mutations in familial NCCM

Hoedemaekers et al., EHJ, 2007





Could other known cardiomyopathy genes be involved in NCCM ????

Gene mutations in LVNC

Gene	Disease	Location	Child/adult
α-Dystrobrevin (DTNA)	LVNC with CHD, muscular dystrophy in human	18q12	Child
G4.5 (TAZ)	Barth syndrome, LVNC, DCM, EFE	Xq28	Child
LIM domain binding protein (LDB3, Cypher/ZASP)	LVNC, DCM	10q22-q23.2	Child/adult
Lamin A/C	LVNC, DCM, muscular dystrophy in human	1q22	Child/adult
Sarcomere proteins			
β-myosin heavy chain (MYH7)	HCM, DCM, LVNC	14q11.2-q13	Adult
α-cardiac actin (ACTC)	HCM, DCM, LVNC	15q11-q14	Adult
Cardiac troponin T (TNNT2)	HCM, DCM, LVNC	1q32	Adult

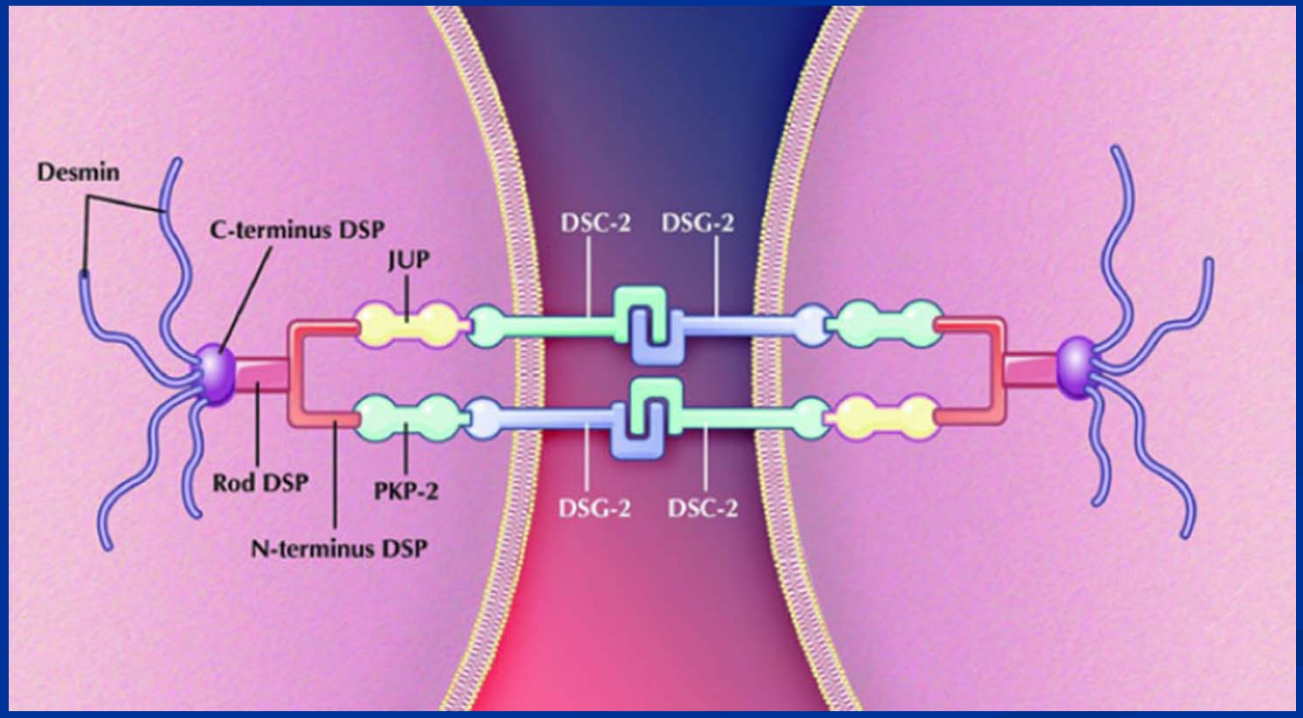
LVNC, left ventricular noncompaction; CHD, congenital heart disease; DCM, dilated cardiomyopathy; EFE, endocardial fibroelastosis; HCM, hypertrophic cardiomyopathy

Prognosis

- In a series of 34 adults with LVNC, 47% either died or underwent cardiac transplantation during the follow-up period of 44±39 months *Oechslin et al. JACC 2000*

- Nearly 60% of patients described in one large series had either died or undergone cardiac transplantation within 6 years of diagnosis *Ritter et al. Mayo Clin Proc. 1997*

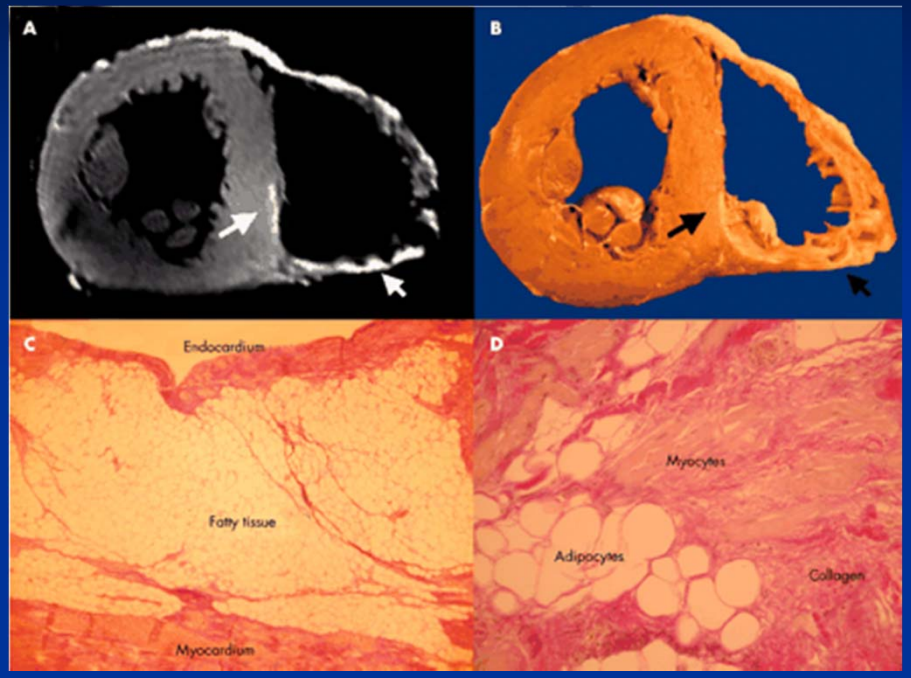
Arrhythmogenic RV Cardiomyopathy

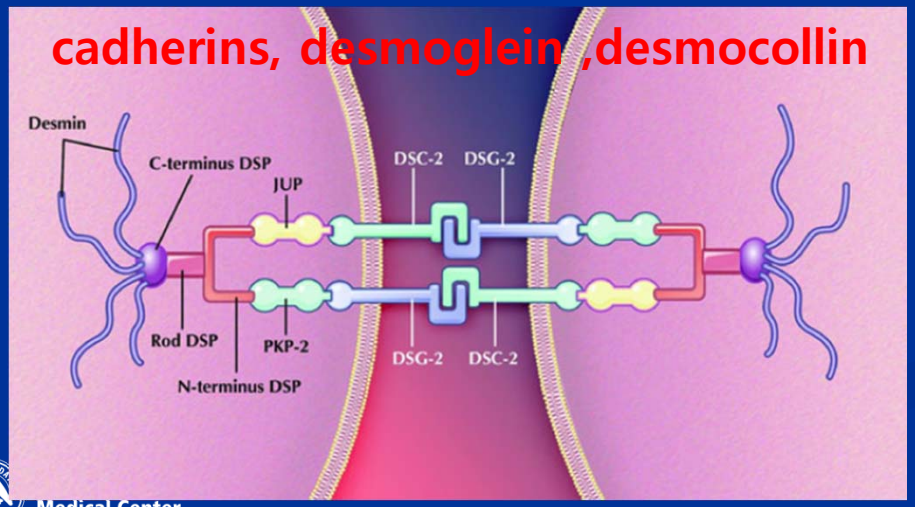


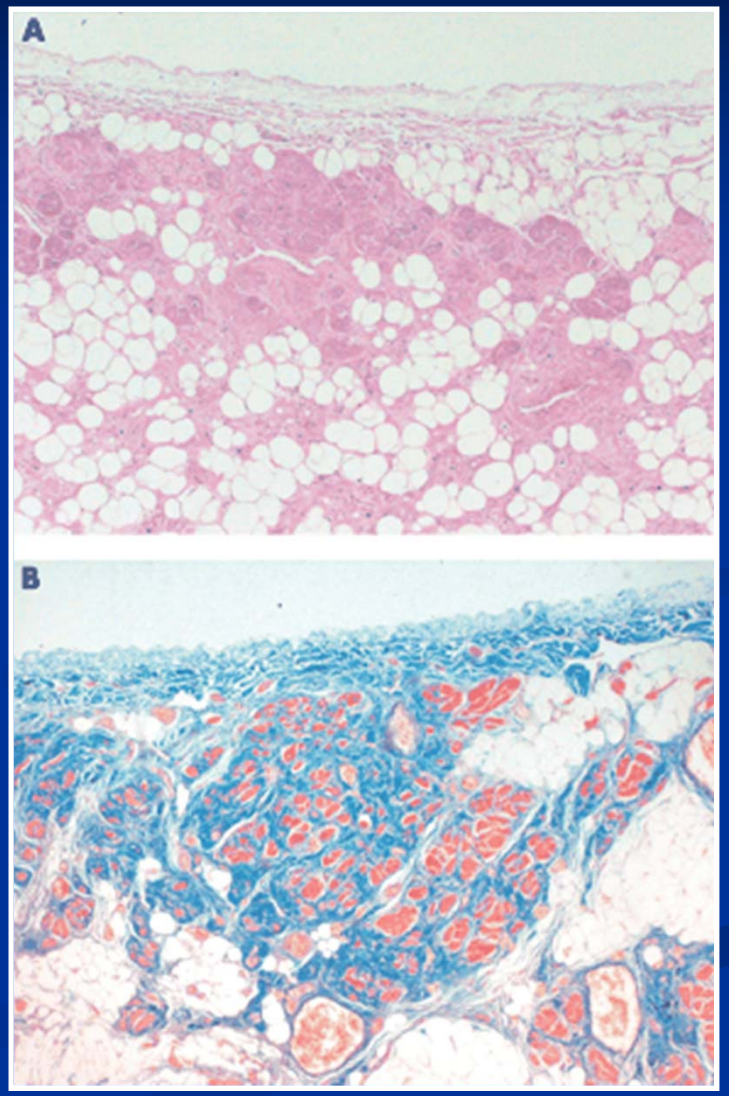
Arrhythmogenic RV dysplasia

- Pathologic hallmark of ARVD is RV myocyte loss with fibrofatty replacement
- Over the past decade, mutations in several desmosomal proteins have been identified as the genetic basis
- Defective desmosomal proteins may lead to impaired mechanical coupling between individual cells, leading to myocyte detachment and degeneration
- RV cavity dilation, ventricular wall thinning with aneurysms, and systolic dysfunction.

Arrhythmogenic RV Dysplasia







Genetic factors

- Comprehensive mutation screening of all known and candidate genes is liable to yield a success rate of at least 40% in ARVD/C
- Lack of an identifiable mutation does not rule out disease
- Mutations were subsequently identified in desmoplakin, desmoglein-2, desmocollin-2, and plakophilin-2 genes
- Several extra-desmosomal genes, such as those encoding TGF-β3, cardiac ryanodine receptor (RyR2), and transmembrane protein 43 (TMEM43), have also been implicated in specific types

Desmosomal Genes Implicated in AVRD/C

Gene (Symbol), Locus	Exons (n), Transcript Size (kb)	Mode of Inheritance	Number of Reported Mutations	Type of Reported Mutations	Associated Phenotype
Plakoglobin (JUP) 17q21	14, 2.4	AR	1	Deletion	Naxos disease
Desmoplakin (DSP) 6p24	24, 8.9	AD AR	>10 3	Various Missense Nonsense Deletion	ARVC ARVC, skin disorder, woolly hair ARVC, skin disorder, woolly hair Carvajal syndrome
Plakophilin (PKP)-2 12p11	14, 2.6	AD AR	>50 1	Various Cryptic splice site	ARVC ARVC
Desmoglein (DSG)-2 18q12	15, 3.4	AD	>20	Various	ARVC
Desmocollin (DSC)-2 18q12	17, 3.1	AD	3	Deletion, insertion, splice site	ARVC

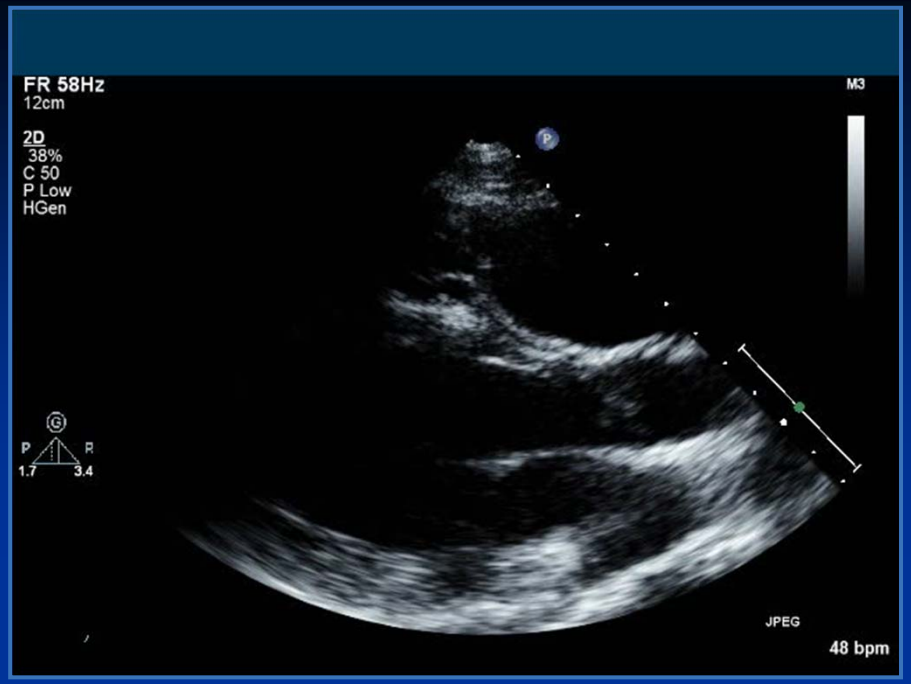
Revised task force criteria for diagnosing arrhythmogenic right ventricular dysplasia

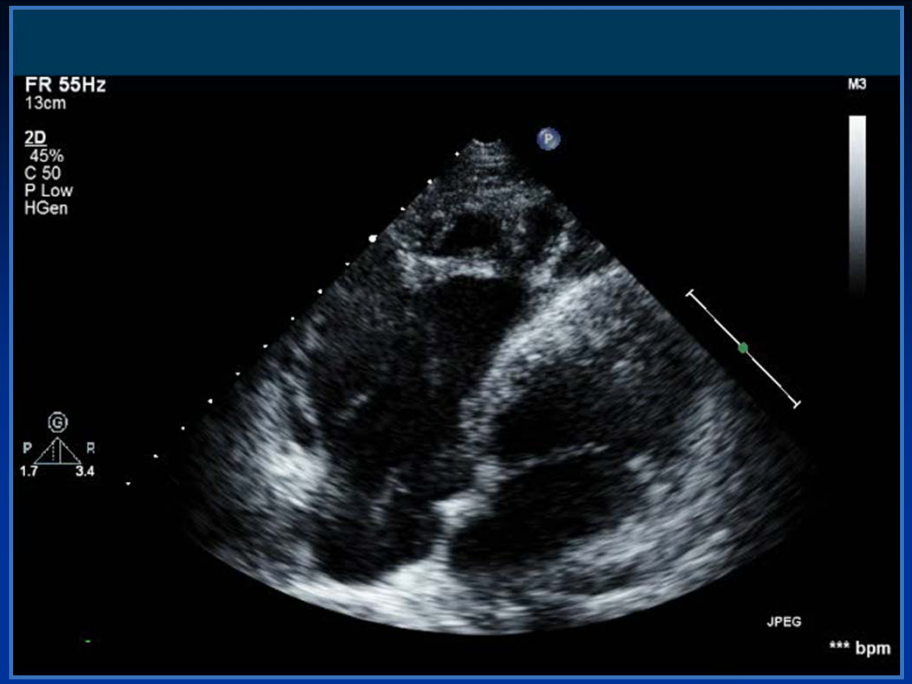
Diagnostic feature	Major criteria	Minor criteria
Global or regional ventricular dysfunction and structural alterations	**By 2D echo:** •Regional RV akinesia, dyskinesia, or aneurysm and 1 of the following (end diastole): -PLAX RVOT ≥32 mm (corrected for body size [PLAX/BSA] ≥19 mm/m2) -PSAX RVOT ≥36 mm (corrected for body size [PSAX/BSA] ≥21 mm/m2) .-or fractional area change ≤33% **By MRI:** •Regional RV akinesia or dyskinesia or dyssynchronous RV contraction •and 1 of the following: -Ratio of RV end-diastolic volume to BSA ≥110 mL/m2 (male) or ≥100 mL/m2 (female) -or RV ejection fraction ≤40% **By RV angiography:** •Regional RV akinesia, dyskinesia, or aneurysm	**By 2D echo:** •Regional RV akinesia or dyskinesia •and 1 of the following (end diastole): -PLAX RVOT ≥29 to G32 mm (corrected for body size [PLAX/BSA] ≥16 to G19 mm/m2) -PSAX RVOT ≥32 to G36 mm (corrected for body size [PSAX/BSA] ≥18 to G21 mm/m2) -or fractional area change >33% to ≤40% **By MRI:** •Regional RV akinesia or dyskinesia or dyssynchronous RV contraction •and 1 of the following: -Ratio of RV end-diastolic volume to BSA ≥100 to G110 mL/m2 (male) or ≥90 to G100 mL/m2 (female) -or RV ejection fraction >40% to ≤45%
Tissue characterization of wall	•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	. Residual myocytes 60% to 75% by morphometric analysis (or 50% to 65% if estimated), with fibrous replacement of the RV free wall myocardium in ≥1 sample, with or without fatty replacement of tissue

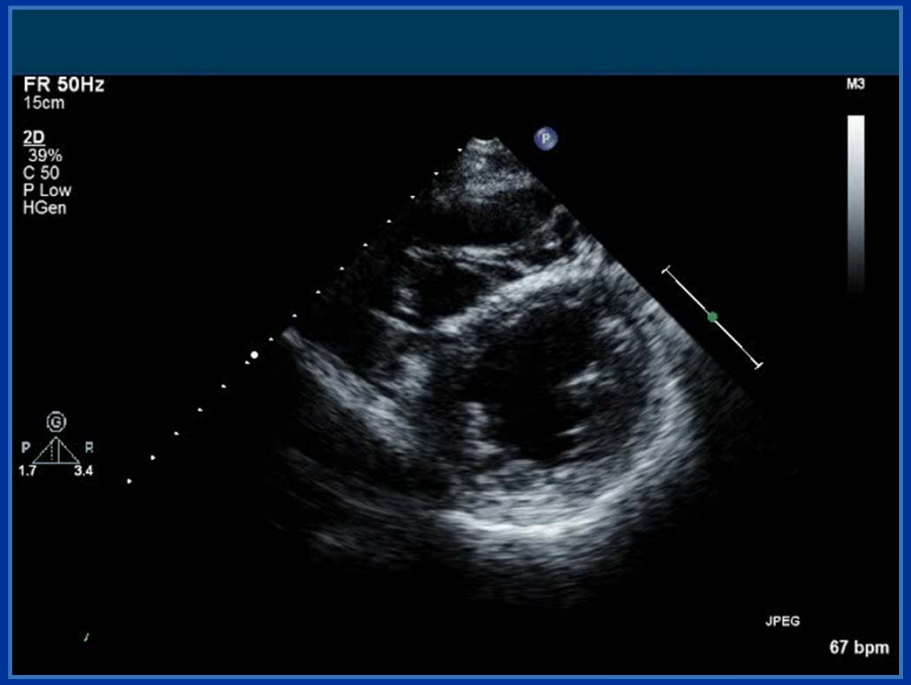
Revised task force criteria for diagnosing arrhythmogenic right ventricular dysplasia

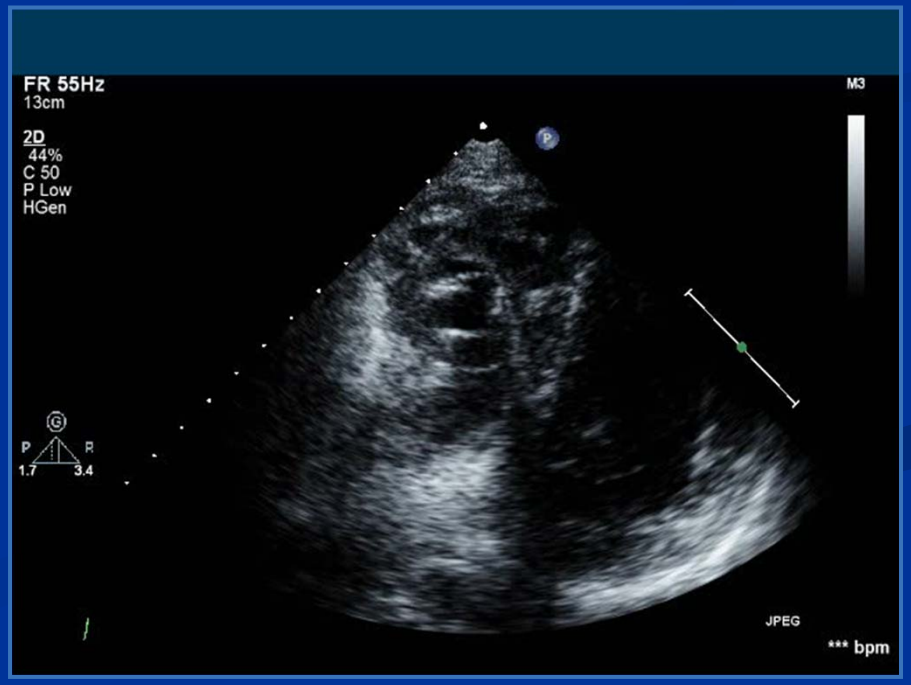
Diagnostic feature	Major criteria	Minor criteria
R… a…	•Inverted T waves in right precordial lea…	•Inverted T waves in leads V1 and V2 in indi-viduals 914 years of age (in the absence of …
D… c… s…	achycardia of LBBB morphology with su perior axis (negative or indeterminate Q RS in leads II, III, and aVF and positive in lead aVL)	end of the QRS, including R',in V1,V2,or V3, in the absenc e of complete RBBB •Nonsustained or sustained ventricular tachy-cardia of RV outflow configuration, LBBB morphology wit h inferior axis (positive QRS in leads II, III, and aVF and n egative in lead aVL) or of unknown axis •>500 ventricular extrasystoles per 24 hours (Holter)

- **The diagnosis of ARVD is based on the presence of two major, one major and two minor, or four minor criteria**
- **Due to the significant heterogeneity in the manifestation of disease, there is no single gold standard test to make the diagnosis of ARVD**
- **Despite being quite specific, these TF criteria lacked sensitivity for diagnosis, especially in presymptomatic patients**


FR 58Hz
12cm
2D
38%
C 50
P Low
HGen
M3
P
G
P R
1.7 3.4
JPEG
48 bpm


FR 55Hz
13cm
2D
45%
C 50
P Low
HGen
M3
P
G
P R
1.7 3.4
JPEG
*** bpm


FR 50Hz
15cm
2D
39%
C 50
P Low
HGen
M3
P
G
P R
1.7 3.4
JPEG
67 bpm


FR 55Hz
13cm
2D
44%
C 50
P Low
HGen
M3
P
G
P R
1.7 3.4
JPEG
*** bpm

Clinical Manifestations

- Palpitations: 67%
- Syncope: 32%
- Atypical chest pain: 27%
- Dyspnea: 11%
- Approximately 50% of patients with ARVD present with ventricular arrhythmias
- Most commonly sustained or nonsustained ventricular tachycardia that originates in the RV, therefore has a LBBB pattern if captured on EKG.
- VT can be exercised induced in patients with ARVD

Treatment

- Major goal is suppression of ventricular tachyarrhythmias (beta blockers)
- Antiarryhthmic medications (sotalol is usually first line agent)
- Avoid competitive sports
- Radiofrequency ablation
- European Society of Cardiology gave ICD placement class I recommendation for secondary prevention, class II a for primary prevention

Stress-Induced Cardiomyopathy

Classification (3)

Cardiomyopathies

HCM

Familial
- Sacomere protein mutation
- Storage disease
- Disorders of fatty acid metabolism
- Carnitine deficiency
- Phosphorylase B kinase deficiency
- Mitochondrial cytopathies
- Syndromic HCM
- Ohters

Non-familial
- Obesity
- Infants of diabetic mothers
- Athletic training
- Amyloid (AL/prealbumin)

DCM

Familial
- Sacomere protein mutation
- Z-band
- Cytoskeletal genes
- Nuclear membrane
- Mildly dilated CM
- Intercalated disc protein mutations (see ARVC)
- Mitochondrial cytopathy

Non-familial
- Myocarditis
- Kawasaki disease
- Eosinophilic (Churg Strauss syndrome)
- Viral persistence
- Drugs
- Pregnancy
- Endocrine
- Nutritional
- Alcohol
- Tachycardio myopathy

ARVC

Familial
- Intercalated disc protein mutations
- Cardiac ryanodine receptor (RyR2)
- Transforming growth factor-β3 (TGFβ3)

Non-familial
- Inflammation

RCM

Familial
- Sarcomeric protein mutations
- Familial amyloidosis
- Desminopathy
- Pseuxanthoma elasticum
- Haemochromatosis
- Anderson-Fabry disease
- Glycogen storage disease

Non-familial
- Amyloid (AL/prealbumin)
- Scleroderma
- Endomyocardial fibrosis
- Carcinoid heart disease
- Metastatic cancers
- Radiation
- Drugs (anthracyclines)

Unclassified

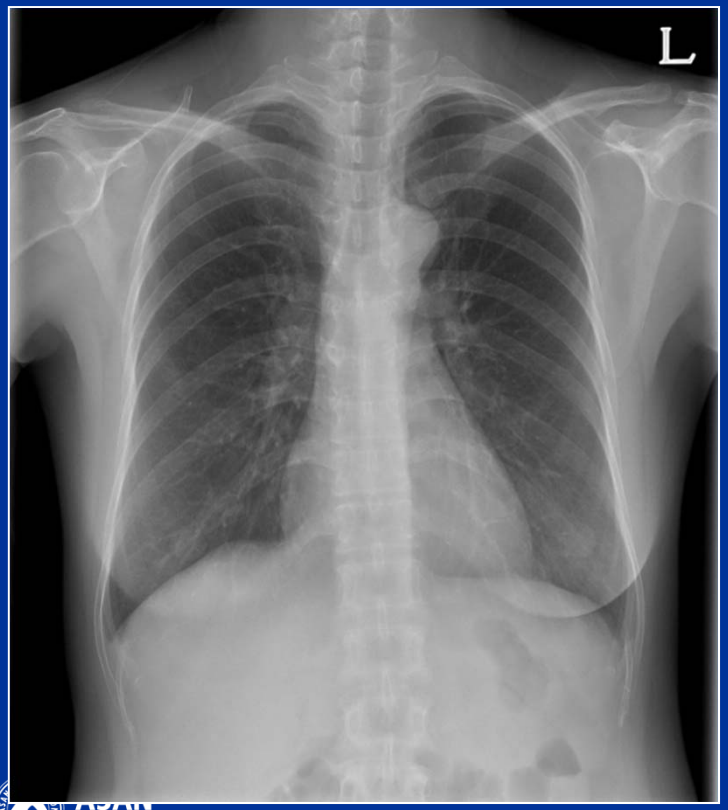
Familial
- Left ventricular non-compaction
  - Barth syndrome
  - Lamin A/C
  - ZASP
  - α-dystrobrevin

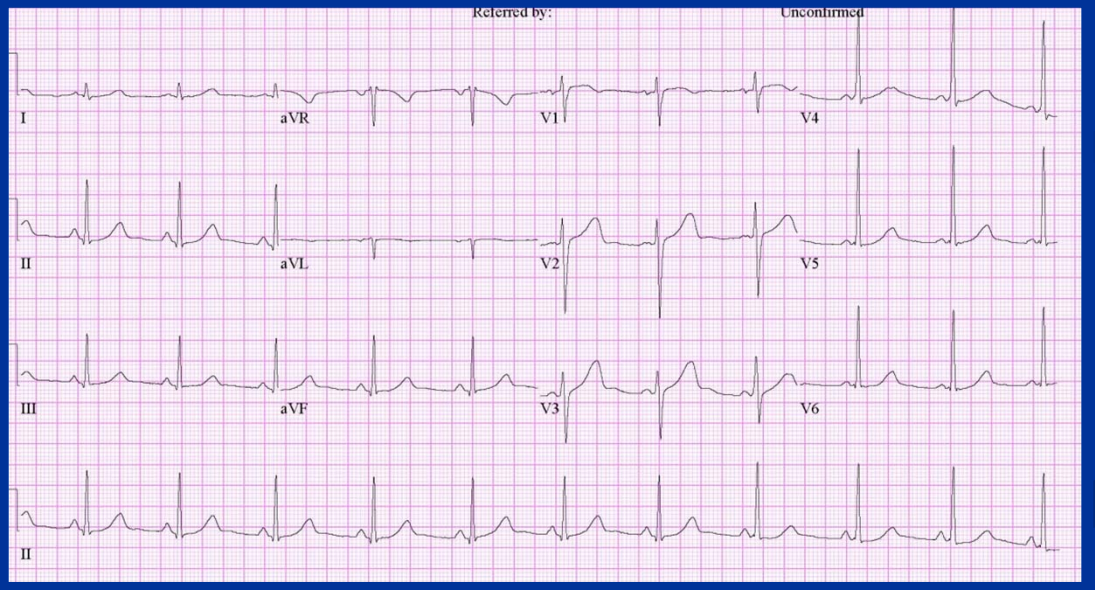
Non-familial
- Tako Tsubo cardiomyopathy

2008 ESC recommendation

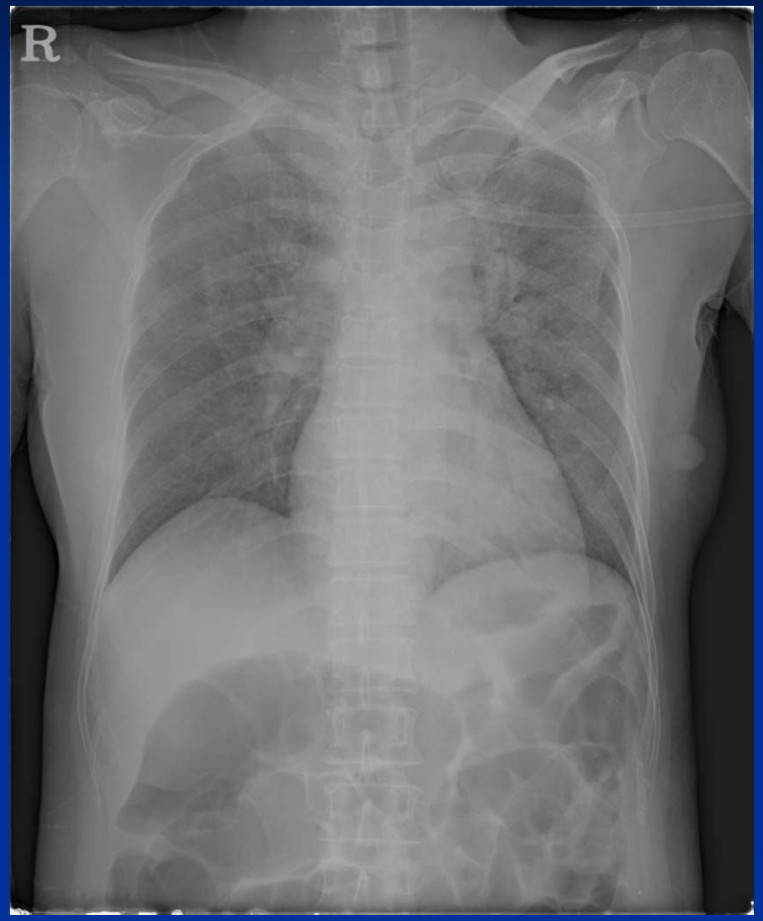
Case

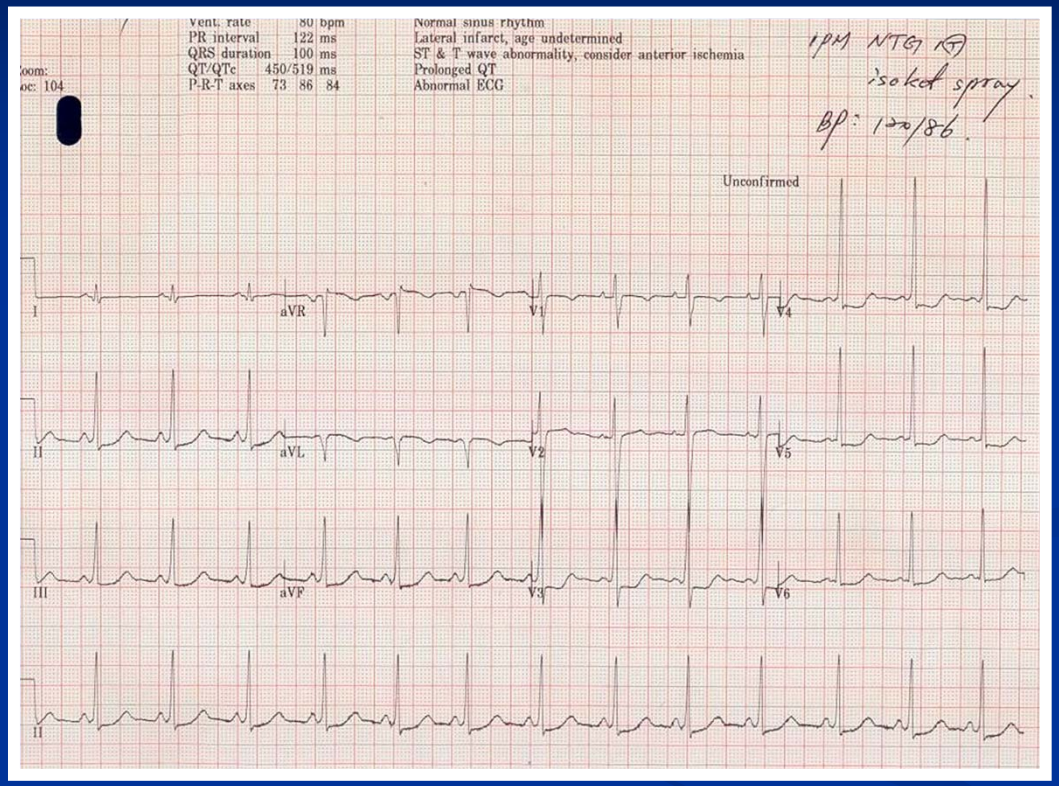
- F/49
- She underwent diagnostic coronary angiography two times due to chest pain, which showed no significant lesion two years ago.
- Sudden dyspnea was developed during routine annual colonofiberscopy in July 2009





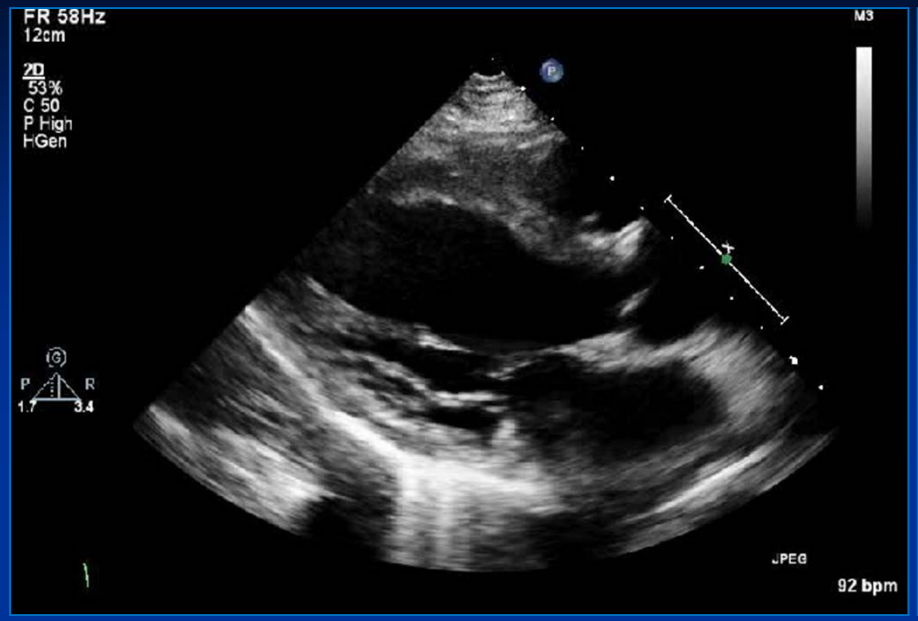
Emergency Department

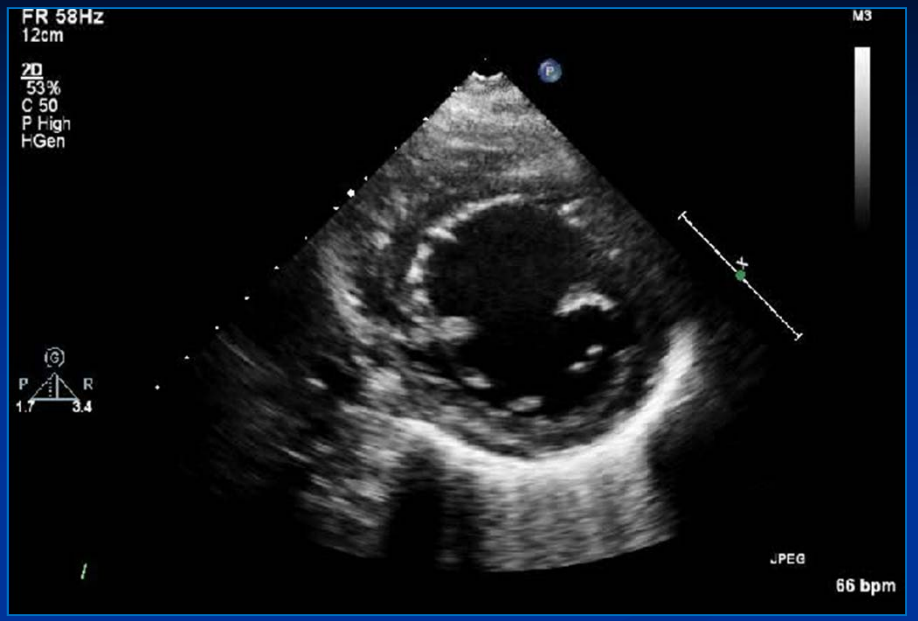


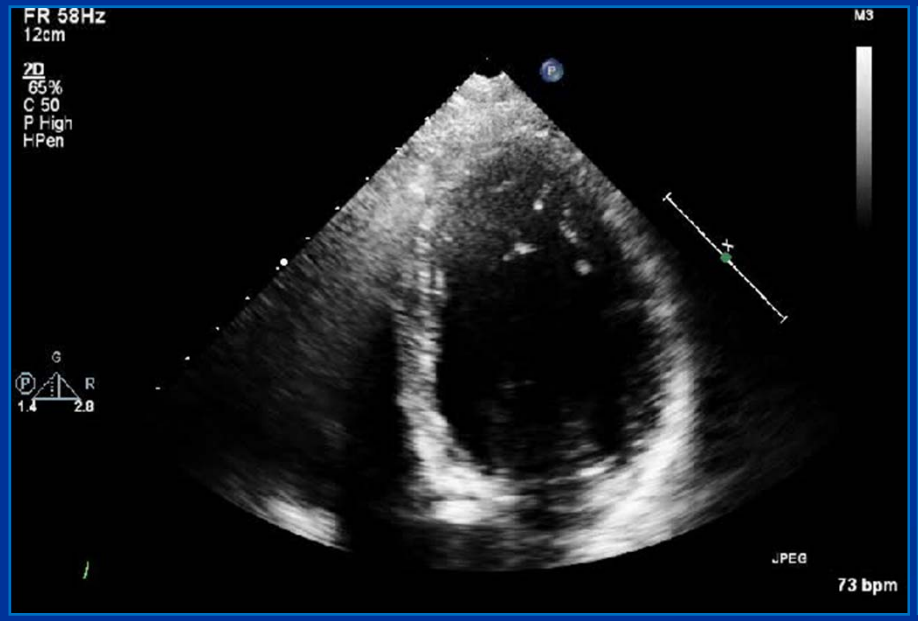


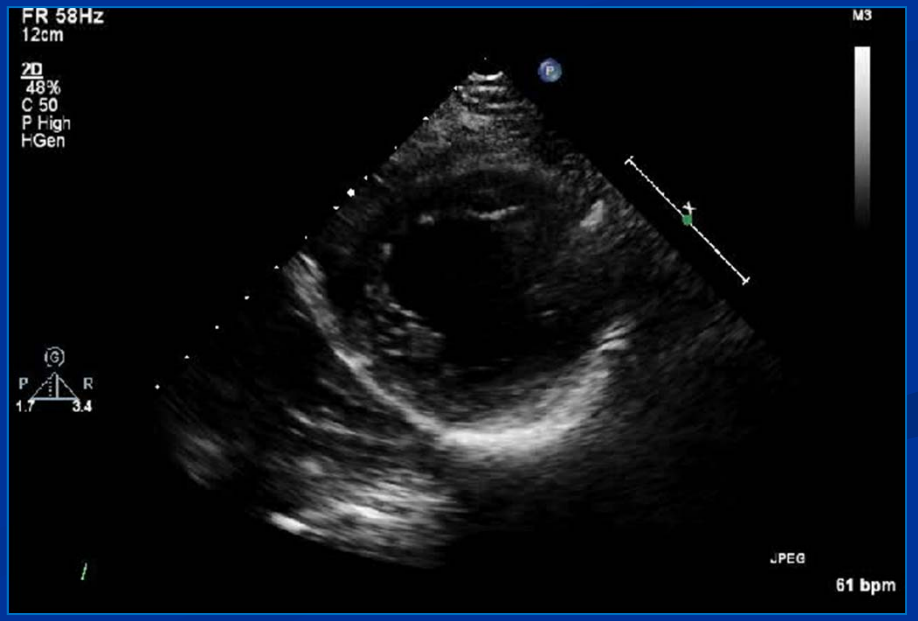
BP 145/84 mmHg, HR 83 bpm

Peak CK-MB/troponin- I = 21.3/2.5 ng/mL

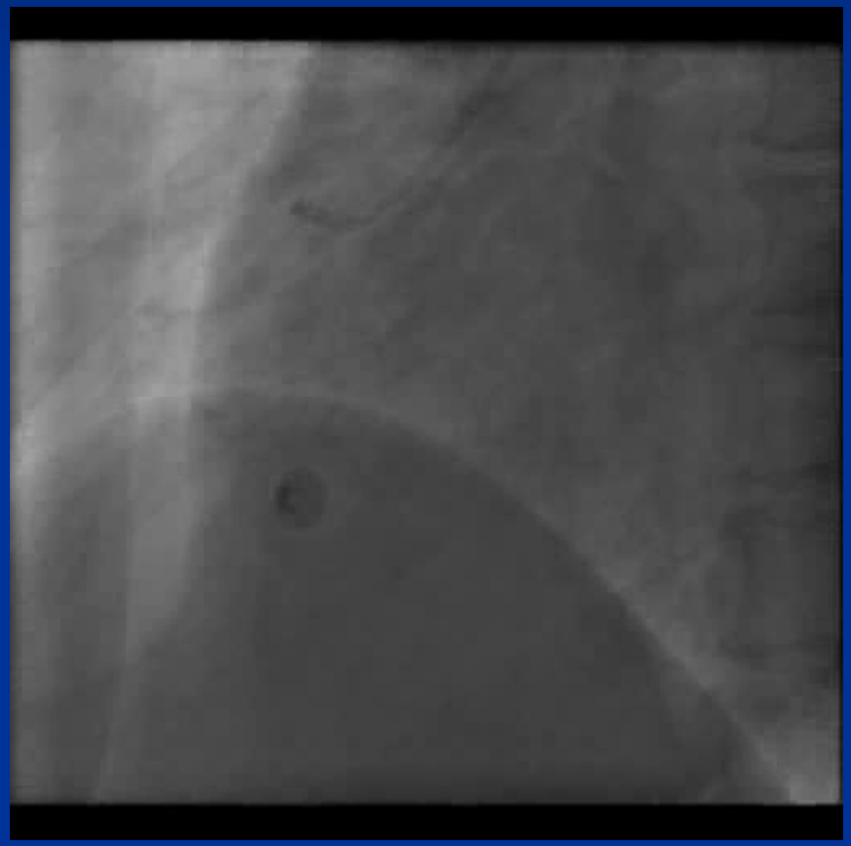


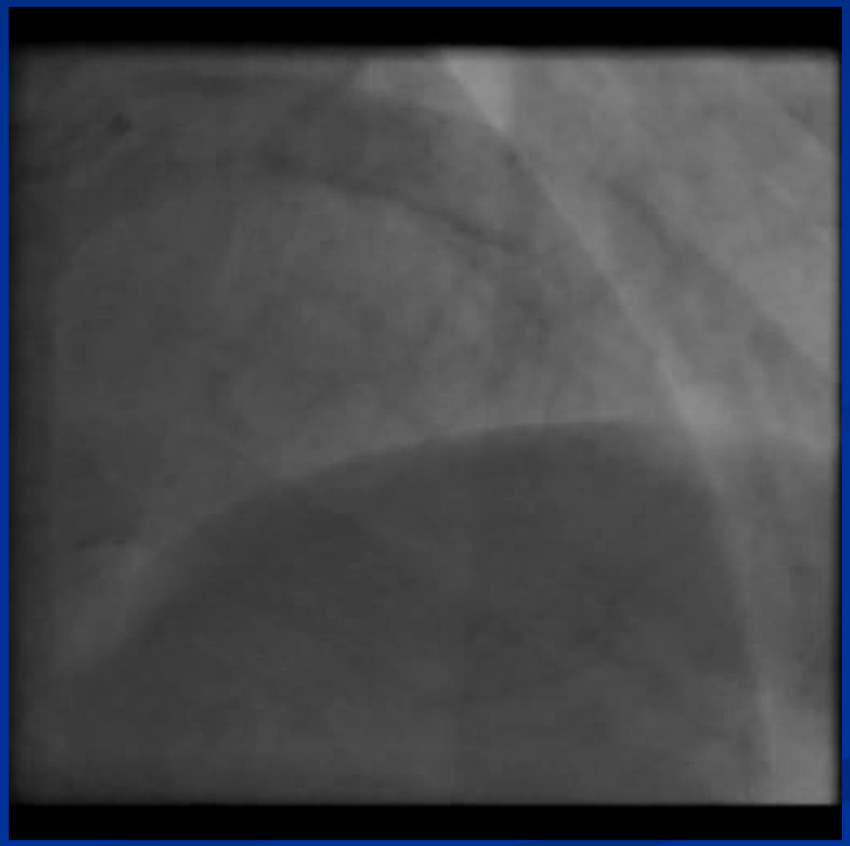


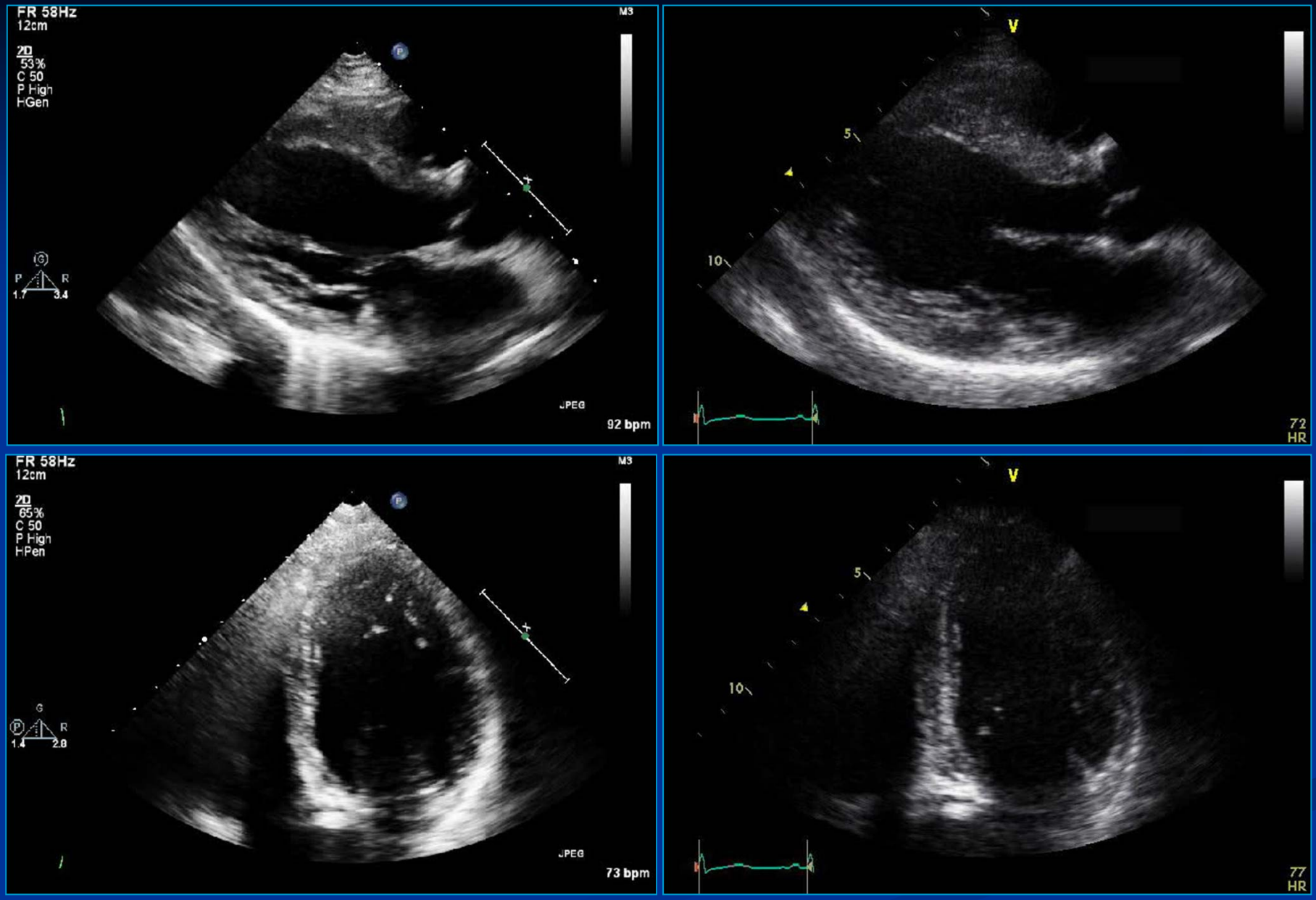




CAG findings



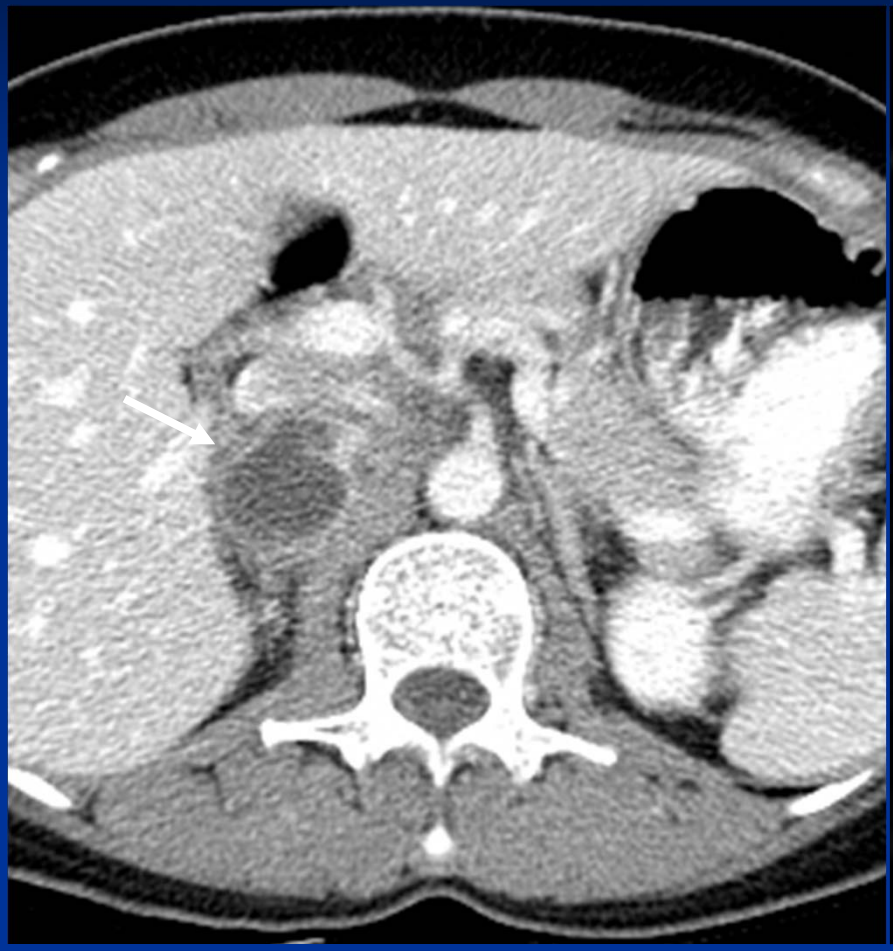




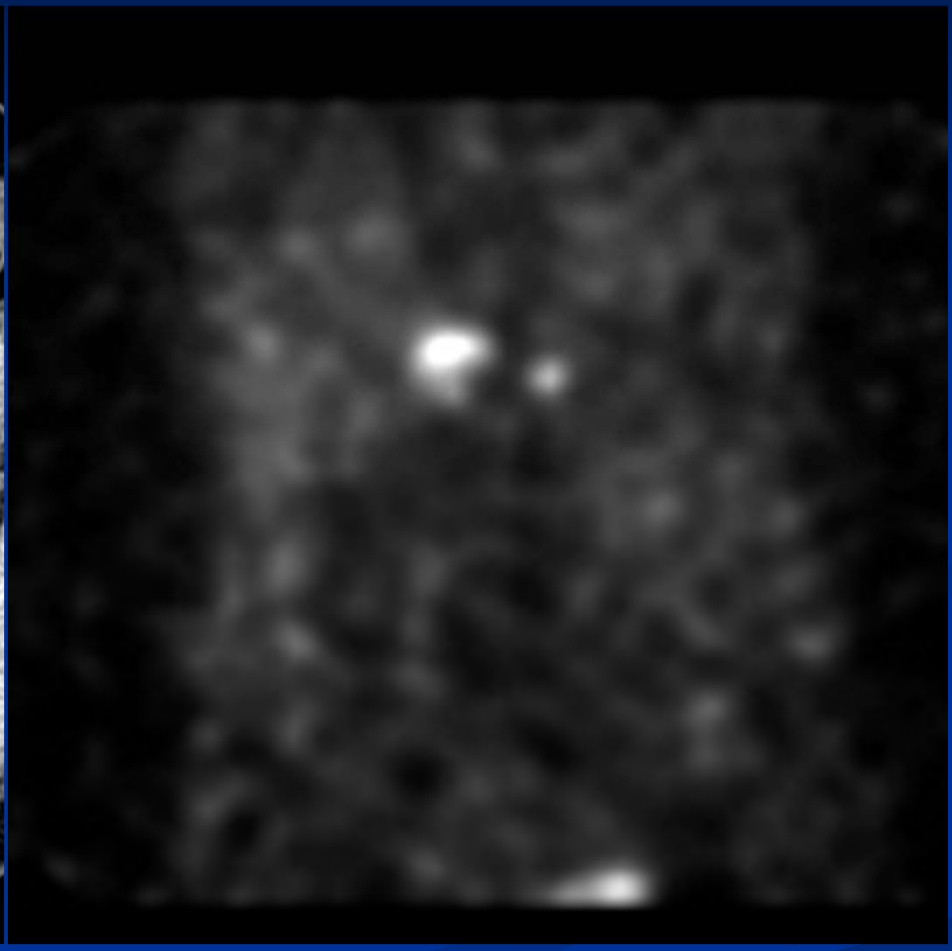
2009.7.24.

2009.7.28.

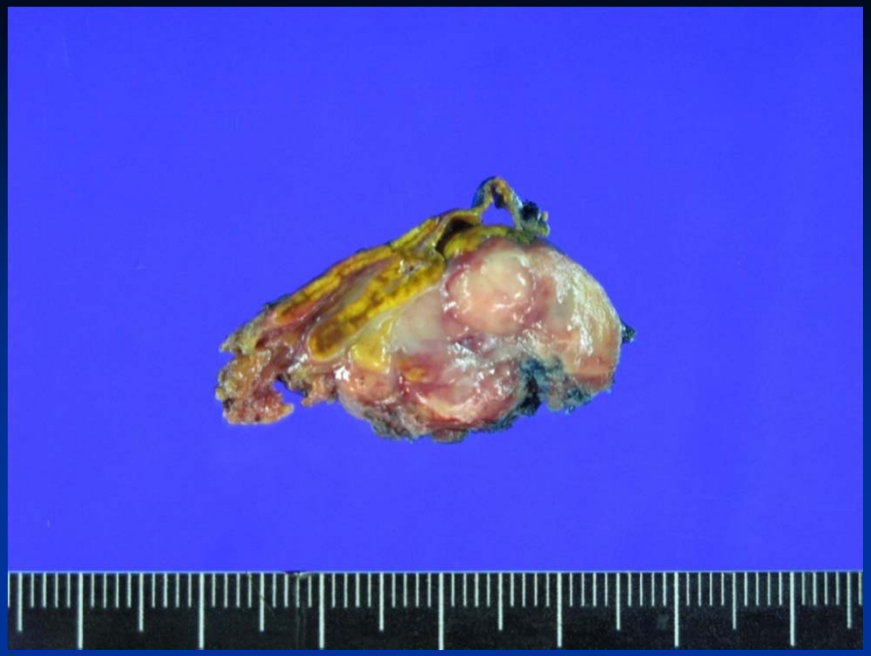
Adrenal Mass

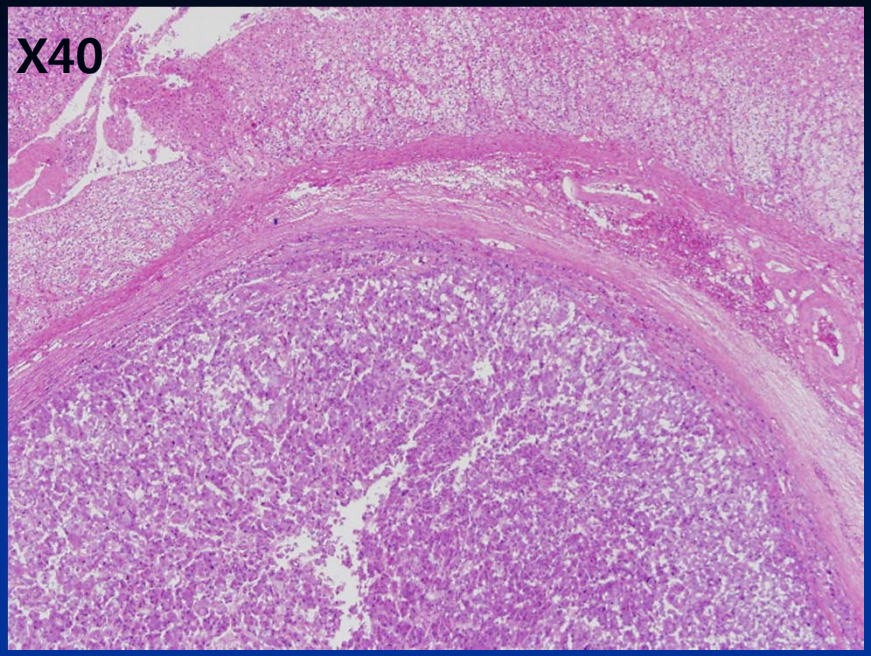


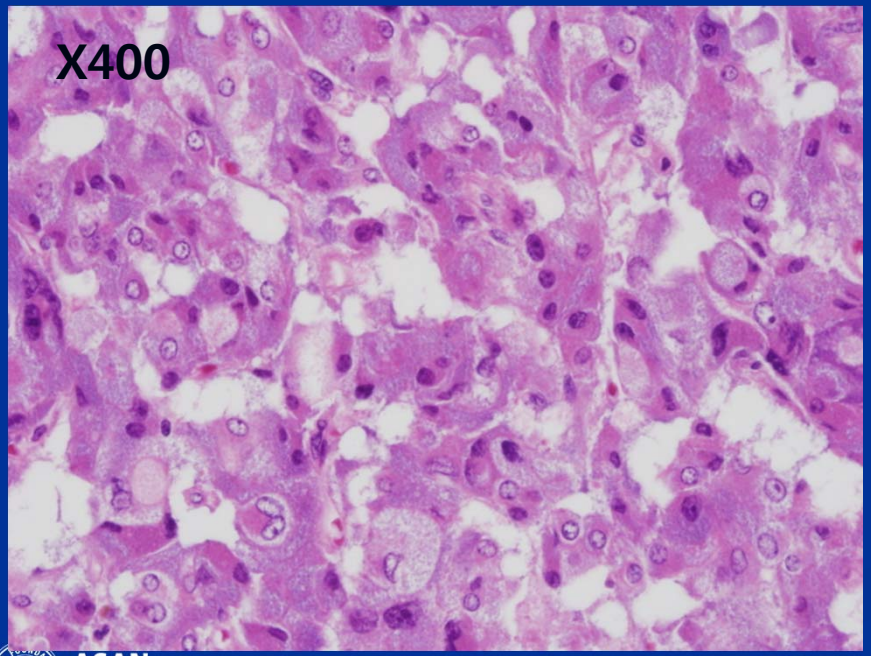
CT

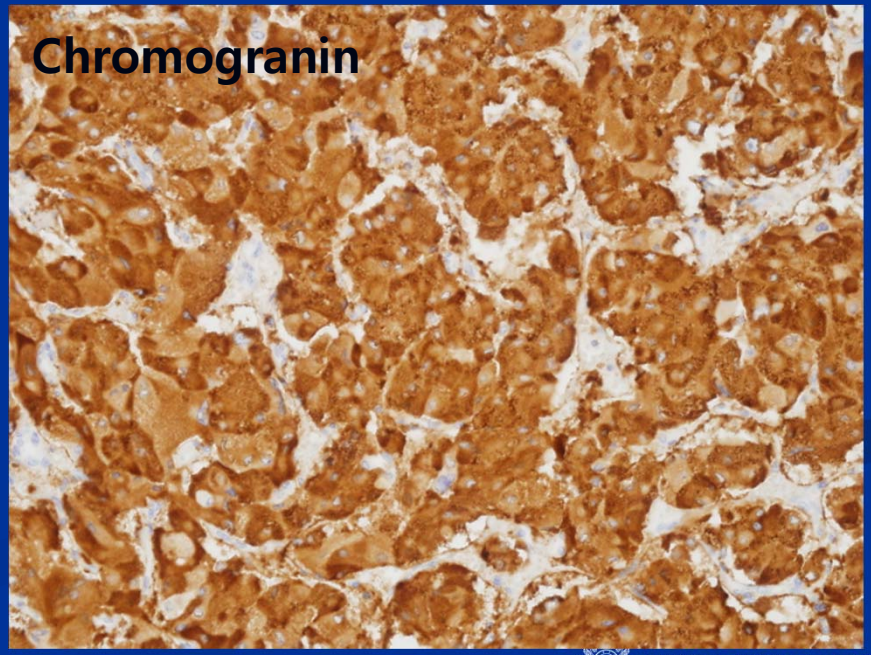


I-123 MIBG scan

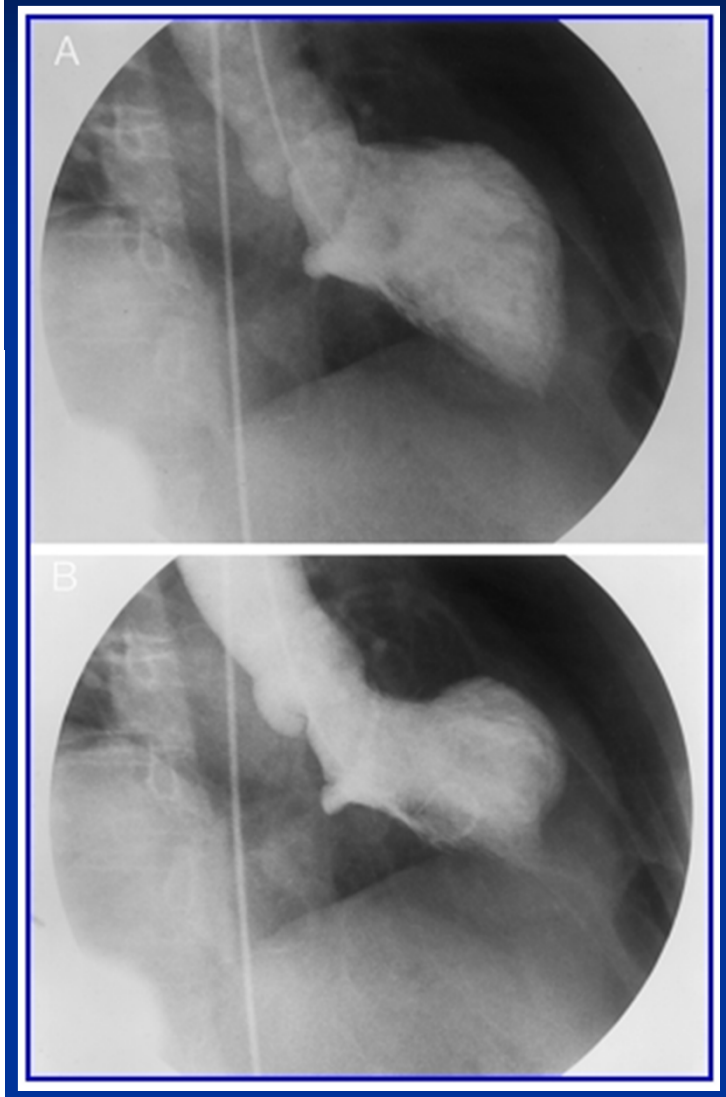



X40


X400

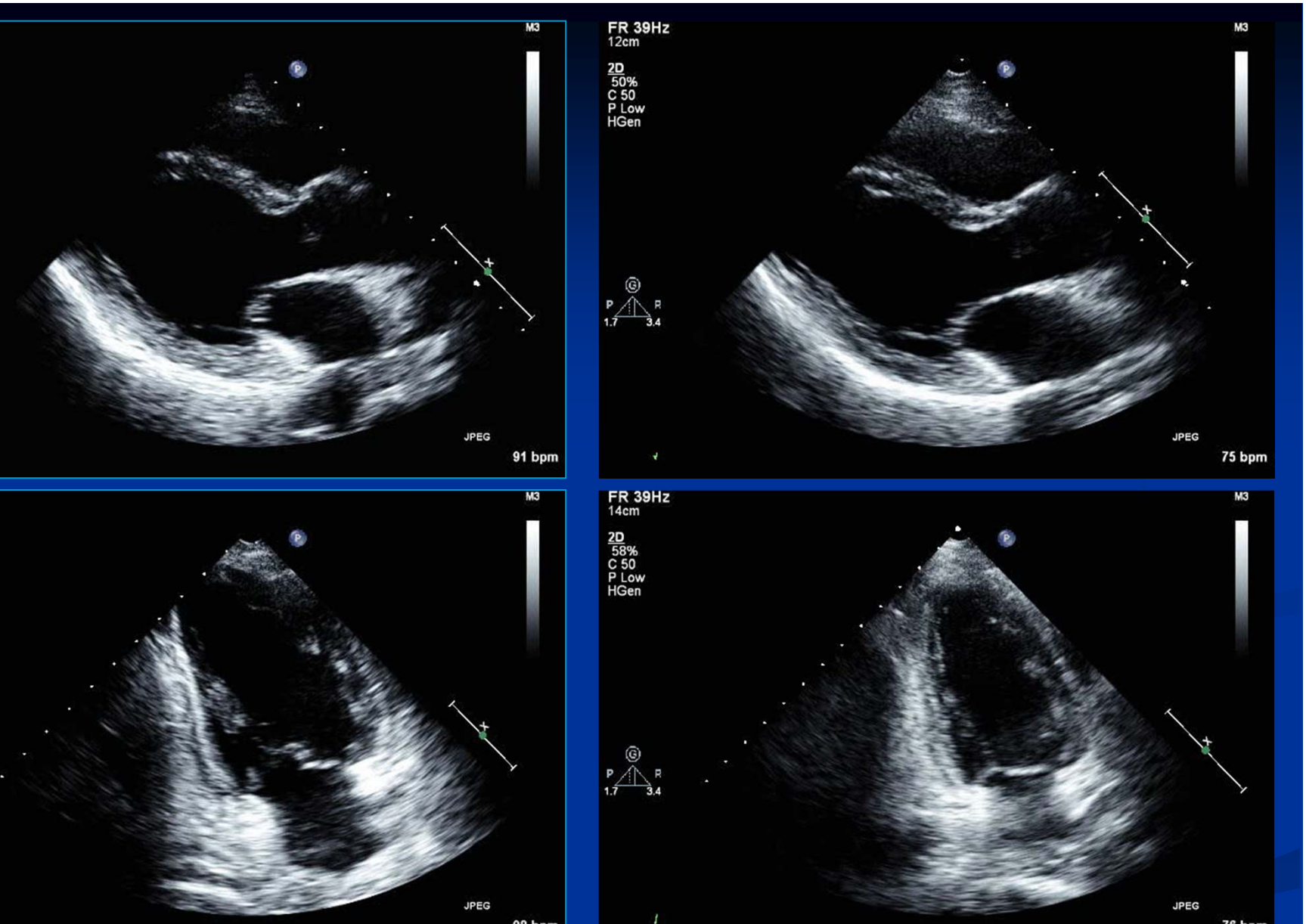

Chromogranin

A Broken Heart



- A 70 yo female without past medical Hx
- Near collapse and chest pain that occurred when she was informed that her husband of 45 years had died of heart failure and refractory ventricular arrhythmias
- ECG; minor anterior ST segment elevations and inferolateral ST depressions, which resolved after IV NTG
- Normal CAG
- Echo 3 months later was normal

Circulation 1998;98:1349


M3
JPEG
91 bpm
FR 39Hz
12cm
2D
50%
C 50
P Low
HGen
P
R
1.7
3.4
M3
JPEG
75 bpm
M3
JPEG
FR 39Hz
14cm
2D
58%
C 50
P Low
HGen
P
R
1.7
3.4
M3
JPEG

Stress-Induced Cardiomyopathy

- **Early investigators**

Postmenopausal women, emotional stress
Differential diagnosis of ACS or NSTEMI
Excellent prognosis

- **Physical stress as an important precipitant**

acute medical illness; sepsis, hypoxemia
surgery/procedures; sigmoidoscopy, ERCP, bronchoscpopy, uneventful GI or cardiac surgery

- **Dyspnea, hypotension, ECG abnormality, O2 desaturation**
- **High mortality with sudden cardiac death and recurrence**

Stress-Induced Cardiomyopathy
: Mechanism

Coronary micro-circulation

Multi-vessel spasm

Catecholamine mediated cardiotoxicity

a Catecholamine and Neuropeptide Levels.*

	Patients with Stress Cardiomyopathy (N=13)			Patients with Killip Class III Myocardial Infarction (N=7)			Normal Value
	Day 1 or 2	Day 3, 4, or 5	Day 7, 8, or 9	Day 1 or 2	Day 3, 4, or 5	Day 7, 8, or 9	
	median (interquartile range)						
e precursor (pg/ml)							
amines (pg/ml)							
ne	1264 (916–1374)†	1044 (733–1118)†	348 (180–550)	376 (275–476)	330 (220–385)	275 (220–311)	37‡
phrine	2284 (1709–2910)†	1573 (1235–2589)†	1142 (525–1252)	1100 (914–1320)	829 (727–914)	541 (516–660)	169‡
e	111 (106–146)†	77 (63–110)	56 (47–77)	61 (46–77)	61 (61–77)	38 (30–61)	15‡
ylacetic acid	2758 (2573–3077)	2598 (2354–2892)†	1345 (1194–1682)	1513 (1211–1648)	1228 (1026–1362)	1009 (908–1059)	1497‡
metabolites (pg/ml)							
	178 (140–187)	509 (385–789)	659 (590–738)§	106 (89–124)	203 (177–213)	205 (189–243)	59‡
rine	216 (130–319)	456 (229–569)	661 (551–696)§	160 (145–170)	196 (181–209)	271 (225–288)	55‡
nl)							
Y	186 (162–236)§	185 (158–214)†	136 (90–182)§	77 (60–90)	69 (61–71)	60 (40–65)	51¶
ic peptide	1033 (805–1783)§	450 (205–684)	142 (72–236)	264 (192–483)	268 (249–574)	297 (142–419)	10–93‖
metabolite (pg/ml)							

SCMP in ICU Patients

Ventricular Apical Ballooning Due evere Physical Stress in Patients nitted to the Medical ICU*

ong Park, MD; Soo-Jin Kang, MD; Jae-Kwan Song, MD;
uk Kim, MD; Chae Man Lim, MD; Duk-Hyun Kang, MD; and
k Koh, MD

Results: **Of the 92 patients, 65 (71%) were men, and they had a mean (± SD) age of 63 ± 11 years. LVAB was observed in 26 patients (28%), with a mean lowest ejection fraction of 33 ± 8% (range, 19 to 46%). Compared with the 66 patients (72%) without LVAB, those with LVAB had a higher frequency of sepsis (62% vs 14%, respectively; $p < 0.001$), a higher prevalence of hypotension on ICU admission, more frequent use of inotropic agents, and a higher frequency of cardiomegaly and pulmonary edema ($p < 0.005$ for each). Sepsis was the only variable associated with the development of LVAB (odds ratio, 9.2; 95% confidence interval, 2.4 to 35.8; $p < 0.001$). The development of Q-wave or ST-segment displacement was associated with LVAB, but the sensitivities were 12% and 19%, respectively. Serum creatine kinase level was elevated in 12 of 26 patients (46%) with LVAB. The normalization of this condition occurred in 20 of 26 patients (77%) a mean duration of 7.4 ± 5.6 days later (range, 2 to 25 days). The mean 2-month survival rate was lower in patients with LVAB than in those without (71 ± 6% vs 52 ± 10%, respectively;**

Apical Ballooning in MICU: Results

	AB (-) (n=66)	AB (+) (n=26)	P value
	63.3 ± 12	63.3 ± 10	0.968
sex	48 (73%)	17 (65%)	0.612
factor for atherosclerosis			
pertension	24 (36%)	4 (15%)	0.077
noking	29 (44%)	13 (50%)	0.647
abetes	17 (26%)	9 (35%)	0.445
son for intensive care			<0.001
poxia and respiratory failure	36 (55%)	10 (39%)	
psis	9 (14%)	16 (61%)	
ners	15 (31%)	0 (0%)	

SCMP in a Tertiary Referral Hospital

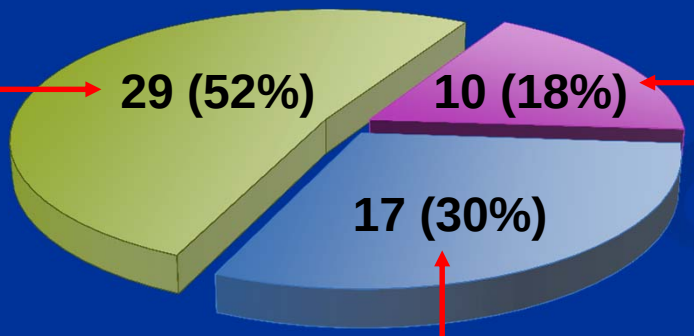
Outcomes of Patients with Stress-Induced Cardiomyopathy Diagnosed by Echocardiography in a Tertiary Referral Hospital

Hyung Lee, MD, Jae-Kwan Song, MD, Byung Joo Sun, MD, Hyung Oh Choi, MD, Jeong-Sook Seo, MD, Jin Oh Na, MD, Dae-Hee Kim, MD, Jong-Min Song, MD, Duk-Hyun Kang, MD, Jae-Joong Kim, MD, and Seong-Wook Park, MD, *Seoul, South Korea*

Over a 63-month period, 56 consecutive patients (median age and interquartile range = 64 years ears]) were identified. Women comprised 79% (44/56) of all patients. The triggering events were acute illness, including sepsis and hypoxemia in 29 patients (52%, group A), in-hospital surgery/procedure tients (30%, group B), and emotional stress in 10 patients (18%, group C). Chest pain was more fre- observed in group C (50%) than in groups A (14%) and B (6%) (P = .021), whereas dyspnea was the ng symptom in groups A and B. Typical takotsubo and inverted takotsubo were observed in 48 and 8 , with a median ejection fraction of 33%. Other abnormalities included dynamic LV outflow tract ion (n = 2), LV thrombus (n = 2), and right ventricular dysfunction (n = 12). Nine deaths (16%) occurred ospitalization. The groups did not differ in mortality. The Acute Physiology and Chronic Health Eval- score (odds ratio 1.405; 95% confidence interval, 1.091-1.810; P = .009) and absence of LV function within 1 week (ejection fraction < 50%) (odds ratio 14.080; 95% confidence interval, 1.184-167.475;) were independent factors associated with mortality. During clinical follow-up up to 6 months, 3 more died, 2 of whom had recurrences of SIC.

SCMP in a Tertiary Referral Hospital

56 consecutive patients in AMC
Median age 64 years (52 – 74)
Female 44 patients (79%)
Three triggering events



Emotional

- argument (3)
- death of family number (3)
- financial loss (2)

Procedure/Surgery

- surgery (11)
- sigmoidoscopy (1)

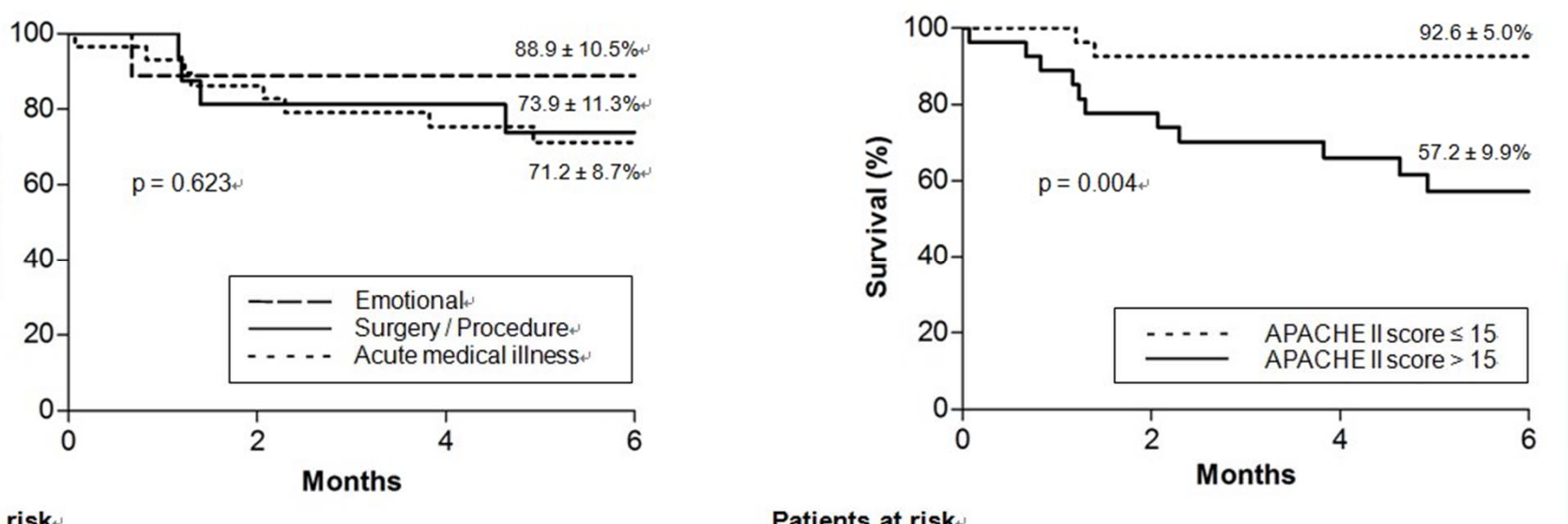
Acute medical illness

- sepsis (15)

SCMP in a Tertiary Referral Hospital

6M mortality 13/56 (23%)

Associated factors: RV dysfucntion & APACHE score



Summary

Classification of cardiomyopathy

- 1995 WHO classification
- 2008 ESC classification
- Will be continued.....

Noncompaction cardiomyopathy

- NCCM shares disease genes and pathophysiological mechanisms with DCM and HCM
- Diagnostic criteria of NCCM
- Comorbid conditions

Summary

ARVC

- Diagnostic criteria
- Genetics
- Risk stratification & ICD insertion

SCMP

- Unknown etiology
- Variable clinical situation
- Multiple RWMAs mimicking ACS
- Variable clinical course !!

Thank you for attention !!!