Non-compaction & Other Unusual Cardiomyopathies

Dae-Hee Kim, MD, PhD
College of Medicine
University of Ulsan
Asan Medical Center
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
  • Sarcomere protein mutation
  • Storage disease
  • Disorders of fatty acid metabolism
  • Carnitine deficiency
  • Mitochondrial cytopathies
  • Syndromic HCM
  • Others
- Non-familial
  • Obesity
  • Infants of diabetic mothers
  • Athletic training
  • Amyloid (AL/prealbumin)

DCM
- Familial
  • Sarcomere 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)
  • Left ventricular non-compaction
  • Barth syndrome
  • Lamin A/C
  • ZASP
  • α-dystrobrevin
- Non-familial
  • Tako Tsubo cardiomyopathy

RCM
- Familial
  • Sarcomeric protein mutations
  • Familial amyloidosis
  • Desminopathy
  • Pseudoxanthoma 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

<table>
<thead>
<tr>
<th></th>
<th>Pediatric patients</th>
<th>Adult patients</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Ichida [21]</td>
<td>Oechslin [31]</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sengupta [36]</td>
</tr>
<tr>
<td>LV apex</td>
<td>100%</td>
<td>94%</td>
</tr>
<tr>
<td>LV inferior wall</td>
<td>70%</td>
<td>94%</td>
</tr>
<tr>
<td>LV lateral wall</td>
<td>41%</td>
<td>100%</td>
</tr>
<tr>
<td>Basal segments</td>
<td></td>
<td></td>
</tr>
<tr>
<td>of the LV wall</td>
<td>&lt; 20%</td>
<td>&lt; 27%</td>
</tr>
</tbody>
</table>
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
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
- Arrhythmias: VT, AF

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Pediatric patients</th>
<th>Adult patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>8</td>
<td>27</td>
</tr>
<tr>
<td>Age at diagnosis (median)</td>
<td>7 yrs</td>
<td>5 yrs</td>
</tr>
<tr>
<td>Male</td>
<td>63%</td>
<td>56%</td>
</tr>
<tr>
<td>Familial occurrence</td>
<td>50%</td>
<td>44%</td>
</tr>
<tr>
<td>Follow-up</td>
<td>up to 5 yrs</td>
<td>up to 17 yrs</td>
</tr>
<tr>
<td>ECG</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bundle branch block</td>
<td>25%</td>
<td>15%</td>
</tr>
<tr>
<td>WPW-syndrome</td>
<td>13%</td>
<td>15%</td>
</tr>
<tr>
<td>Ventricular tachycardia</td>
<td>38%</td>
<td>0%</td>
</tr>
<tr>
<td>Clinical symptoms</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Heart failure symptoms</td>
<td>63%</td>
<td>30%</td>
</tr>
<tr>
<td>Systemic embolic events</td>
<td>38%</td>
<td>0%</td>
</tr>
<tr>
<td>Pulmonary embolism</td>
<td>0%</td>
<td>7%</td>
</tr>
<tr>
<td>Ventricular thrombi</td>
<td>25%</td>
<td>0%</td>
</tr>
<tr>
<td>Facial dysmorphism</td>
<td>38%</td>
<td>33%</td>
</tr>
<tr>
<td>Neuromuscular disorders</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Deaths</td>
<td>38%</td>
<td>7%</td>
</tr>
<tr>
<td>Heart transplantation</td>
<td>0%</td>
<td>4%</td>
</tr>
</tbody>
</table>
MYH7 mutations in familial NCCM

Hoedemaekers et al., EHJ, 2007

Could other known cardiomyopathy genes be involved in NCCM ????
Gene mutations in LVNC

<table>
<thead>
<tr>
<th>Gene</th>
<th>Disease</th>
<th>Location</th>
<th>Child/adult</th>
</tr>
</thead>
<tbody>
<tr>
<td>α-Dystrobrevin (DTNA)</td>
<td>LVNC with CHD, muscular dystrophy in human</td>
<td>18q12</td>
<td>Child</td>
</tr>
<tr>
<td>G4.5 (TAZ)</td>
<td>Barth syndrome, LVNC, DCM, EFE</td>
<td>Xq28</td>
<td>Child</td>
</tr>
<tr>
<td>LIM domain binding protein (LDB3, Cypher/ZASP)</td>
<td>LVNC, DCM</td>
<td>10q22-q23.2</td>
<td>Child/adult</td>
</tr>
<tr>
<td>Lamin A/C</td>
<td>LVNC, DCM, muscular dystrophy in human</td>
<td>1q22</td>
<td>Child/adult</td>
</tr>
</tbody>
</table>

Sarcomere proteins

<table>
<thead>
<tr>
<th>Gene</th>
<th>Disease</th>
<th>Location</th>
<th>Child/adult</th>
</tr>
</thead>
<tbody>
<tr>
<td>β-myosin heavy chain (MYH7)</td>
<td>HCM, DCM, LVNC</td>
<td>14q11.2-q13</td>
<td>Adult</td>
</tr>
<tr>
<td>α-cardiac actin (ACTC)</td>
<td>HCM, DCM, LVNC</td>
<td>15q11-q14</td>
<td>Adult</td>
</tr>
<tr>
<td>Cardiac troponin T (TNNT2)</td>
<td>HCM, DCM, LVNC</td>
<td>1q32</td>
<td>Adult</td>
</tr>
</tbody>
</table>

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.

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

cadherins, desmoglein, desmocollin
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

<table>
<thead>
<tr>
<th>Gene (Symbol), Locus</th>
<th>Exons (n), Transcript Size (kb)</th>
<th>Mode of Inheritance</th>
<th>Number of Reported Mutations</th>
<th>Type of Reported Mutations</th>
<th>Associated Phenotype</th>
</tr>
</thead>
<tbody>
<tr>
<td>Plakoglobin (JUP) 17q21</td>
<td>14, 2.4</td>
<td>AR</td>
<td>1</td>
<td>Deletion</td>
<td>Naxos disease</td>
</tr>
<tr>
<td>Desmoplakin (DSP) 6p24</td>
<td>24, 8.9</td>
<td>AD, AR</td>
<td>&gt;10</td>
<td>Various Missense, Nonsense, Deletion</td>
<td>ARVC, skin disorder, woolly hair, Carvajal syndrome</td>
</tr>
<tr>
<td>Plakophilin (PKP)-2 12p11</td>
<td>14, 2.6</td>
<td>AD, AR</td>
<td>&gt;50</td>
<td>Various Cryptic splice site</td>
<td>ARVC, ARVC</td>
</tr>
<tr>
<td>Desmoglein (DSG)-2 18q12</td>
<td>15, 3.4</td>
<td>AD</td>
<td>&gt;20</td>
<td>Various</td>
<td>ARVC</td>
</tr>
<tr>
<td>Desmocollin (DSC)-2 18q12</td>
<td>17, 3.1</td>
<td>AD</td>
<td>3</td>
<td>Deletion, insertion, splice site</td>
<td>ARVC</td>
</tr>
</tbody>
</table>
Revised task force criteria for diagnosing arrhythmogenic right ventricular dysplasia

<table>
<thead>
<tr>
<th>Diagnostic feature</th>
<th>Major criteria</th>
<th>Minor criteria</th>
</tr>
</thead>
</table>
| Global or regional ventricular dysfunction       | 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 dys-synchronous 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  
| Minor criteria                                  | 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 dys-synchronous 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  
| Minor criteria                                  | • 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

<table>
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<tr>
<th>Diagnostic feature</th>
<th>Major criteria</th>
<th>Minor criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Repolarization abnormalities</td>
<td>• Inverted T waves in right precordial leads V1, V2, and V3 in individuals &gt;14 years of age (in the absence of complete RBBB) or ≥120 ms</td>
<td>• Inverted T waves in leads V1 and V2 in individuals 914 years of age (in the absence of complete RBBB) or in V4, V5, or V6</td>
</tr>
<tr>
<td>Depolarization/conduction abnormalities</td>
<td>• Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right precordial leads (V1 to V3)</td>
<td>• Nonsustained or sustained ventricular tachycardia of LBBB morphology with superior axis (negative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL)</td>
</tr>
<tr>
<td></td>
<td>• Late potentials by SAECG in ≥1 of 3 parameters in the absence of a QRS duration of ≥110 ms on the standard ECG</td>
<td>• The diagnosis of ARVD is based on the presence of two major, one major and two minor, or four minor criteria</td>
</tr>
<tr>
<td></td>
<td>• Filtered QRS duration (fQRS) ≥114 ms</td>
<td></td>
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<tr>
<td></td>
<td>• Duration of terminal QRS ≥40 μV (low-amplitude signal duration) ≥38 ms</td>
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<tr>
<td></td>
<td>• Root mean-square voltage of terminal 40 ms ≤20 μV</td>
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<tr>
<td></td>
<td>• Terminal activation duration of QRS ≥55 ms measured from the nadir of the S wave to the end of the QRS, including R', in V1, V2, or V3, in the absence of complete RBBB</td>
<td>• &gt;500 ventricular extrasystoles per 24 hours (Holter)</td>
</tr>
</tbody>
</table>
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)
- Antiarrythmic 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**

<table>
<thead>
<tr>
<th>Category</th>
<th>Familial</th>
<th>Non-familial</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>HCM</strong></td>
<td>- Sacomere protein mutation</td>
<td>- Obesity</td>
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<td></td>
<td>- Intercalated disc protein mutations (see ARVC)</td>
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<tr>
<td></td>
<td>- Mitochondrial cytopathy</td>
<td></td>
</tr>
<tr>
<td><strong>ARVC</strong></td>
<td>- Intercalated disc protein mutations (see ARVC)</td>
<td>- Inflammation</td>
</tr>
<tr>
<td><strong>RCM</strong></td>
<td>- Sarcomeric protein mutations</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- Familial amyloidosis</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- Desminopathy</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- Pseudoxanthoma elasticum</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- Haemochromatosis</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- Anderson-Fabry disease</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- Glycogen storage disease</td>
<td></td>
</tr>
<tr>
<td><strong>Unclassified</strong></td>
<td>- Left ventricular non-compaction</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- Barth syndrome</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- Lamin A/C</td>
<td></td>
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<tr>
<td></td>
<td>- ZASP</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- α-dystrobrevin</td>
<td></td>
</tr>
</tbody>
</table>

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
BP 145/84 mmHg, HR 83 bpm
Peak CK-MB/troponin- I = 21.3/2.5 ng/mL
CAG findings
Adrenal Mass

CT

I-123 MIBG scan
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
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, bronchoscopy, 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
<table>
<thead>
<tr>
<th>Test</th>
<th>Patients with Stress Cardiomyopathy (N=13)</th>
<th>Patients with Killip Class III Myocardial Infarction (N=7)</th>
<th>Normal Value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Day 1 or 2</td>
<td>Day 3, 4, or 5</td>
<td>Day 7, 8, or 9</td>
</tr>
<tr>
<td>norepinephrine (pg/ml)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>1264 (916–1374)†</td>
<td>1044 (733–1118)†</td>
<td>348 (180–550)</td>
</tr>
<tr>
<td>epinephrine (pg/ml)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>2284 (1709–2910)†</td>
<td>1573 (1233–2589)†</td>
<td>1142 (525–1252)</td>
</tr>
<tr>
<td>angiotensin I (ng/l)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>111 (106–146)†</td>
<td>77 (63–110)</td>
<td>56 (47–77)</td>
</tr>
<tr>
<td>Metabolites (pg/ml)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>norepinephrine</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>2758 (2573–3077)</td>
<td>2598 (2354–2892)†</td>
<td>1345 (1194–1682)</td>
</tr>
<tr>
<td>epinephrine</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>angiotensin I</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>Metabolites (pg/ml)</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>norepinephrine</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>186 (162–236)§</td>
<td>185 (158–214)†</td>
<td>136 (90–182)§</td>
</tr>
<tr>
<td>epinephrine</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>1033 (805–1783)§</td>
<td>450 (205–684)</td>
<td>142 (72–236)</td>
</tr>
</tbody>
</table>
SCMP in ICU Patients

Ventricular Apical Ballooning Due to Severe Physical Stress in Patients Admitted to the Medical ICU*

Song Park, MD; Soo-Jin Kang, MD; Jae-Kwan Song, MD; Suk Kim, MD; Chae Man Lim, MD; Duk-Hyun Kang, MD; and Suk 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

<table>
<thead>
<tr>
<th></th>
<th>AB (-) (n=66)</th>
<th>AB (+) (n=26)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex</td>
<td>48 (73%)</td>
<td>17 (65%)</td>
<td>0.612</td>
</tr>
<tr>
<td>Factor for atherosclerosis</td>
<td>24 (36%)</td>
<td>4 (15%)</td>
<td>0.077</td>
</tr>
<tr>
<td>Hypertension</td>
<td>29 (44%)</td>
<td>13 (50%)</td>
<td>0.647</td>
</tr>
<tr>
<td>Smoking</td>
<td>17 (26%)</td>
<td>9 (35%)</td>
<td>0.445</td>
</tr>
<tr>
<td>Diabetes</td>
<td>36 (55%)</td>
<td>10 (39%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Reason for intensive care</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Respiratory failure</td>
<td>36 (55%)</td>
<td>10 (39%)</td>
<td></td>
</tr>
<tr>
<td>Pneumonia</td>
<td>9 (14%)</td>
<td>16 (61%)</td>
<td></td>
</tr>
<tr>
<td>Others</td>
<td>15 (31%)</td>
<td>0 (0%)</td>
<td></td>
</tr>
<tr>
<td>CHF score</td>
<td>68.8 ± 33.4</td>
<td>83.6 ± 33.8</td>
<td>0.0062</td>
</tr>
</tbody>
</table>
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) 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 patients (30%, group B), and emotional stress in 10 patients (18%, group C). Chest pain was more frequently observed in group C (50%) than in groups A (14%) and B (6%) (P = .021), whereas dyspnea was the most frequent symptom in groups A and B. Typical takotsubo and inverted takotsubo were observed in 48 and 8 cases, with a median ejection fraction of 33%. Other abnormalities included dynamic LV outflow tract obstruction (n = 2), LV thrombus (n = 2), and right ventricular dysfunction (n = 12). Nine deaths (16%) occurred prior to hospitalization. The groups did not differ in mortality. The Acute Physiology and Chronic Health Evaluation II scores (odds ratio 1.405; 95% confidence interval, 1.091-1.810; P = .009) and absence of LV function recovery within 1 week (ejection fraction < 50%) (odds ratio 14.080; 95% confidence interval, 1.184-167.475; P = .030) 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

- Acute medical illness
  - sepsis (15)
- Emotional
  - argument (3)
  - death of family number (3)
  - financial loss (2)
- Procedure/Surgery
  - surgery (11)
  - sigmoidoscopy (1)
SCMP in a Tertiary Referral Hospital

6M mortality 13/56 (23%)

Associated factors: RV dysfunction & 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 !!!