

Acute Pulmonary edema
Secondary to
**CARDIAC FAILURE - NON
COMPACTED LEFT
VENTRICLE**
in Pregnancy



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Case summary.....

- Mrs MS, 22yrs, Primi @ 35⁺⁶ weeks
- Diagnosed as preterm labour - Tab Nifedepin 20mgTid + Inj Betamethasone 12 mg IM stat
- Shifted to FH @ 00.15 on 15th October 2013
- State of CV collapse, Non palpable peripheral pulses
- Bradycardia (30 bpm), Tachypnoea (RR 50/min) SpO₂: 77% , O₂ via mask
- Asystole within seconds of arrival to FH
- Immediate resuscitation done
 - CPR / ACLS / ETT / IPPV
- Perimortem CS in HDU @ 00.20 AM
 - Alive male baby, 2.12 kg, Apgar 3/6/8
- ROSC in 4 minutes
 - Extreme persistent tachycardia @ 180/min
- ICU care & Ventilatory Care, Cardiac Monitoring
- IV antibiotics / IV Frusemide / LMWH, Other Supportive measures

Case summary.....

■ PROVISIONAL DIAGNOSIS

- Peripartum CMP Acute CHF
- Underlying HD..... Acute CHF
- TPTL / Depin / Betnesol ... Acute CHF
- Sepsis ALI / ARDS
- Aspiration Pneumonitis ALI / ARDS

Case summary.....

Past Medical History

- H/o admission @ 3 months of age
- s/s of pneumonia / CHD = Diagnosed as CHD
- Was on therapy upto 10 years of age
- No records available
- 2D Echo done pre marriage: NAD
- ? ? VSD Spontaneous closure

Case summary.....

■ INVESTIGATIONS

- CBC, CUE, LFT, RFT , coagulation profile were normal

- 12 Lead ECG

- Sinus Tachycardia

- Inverted T waves (Rate related)

- X-ray Chest

- Cardiomegaly

2D Echo

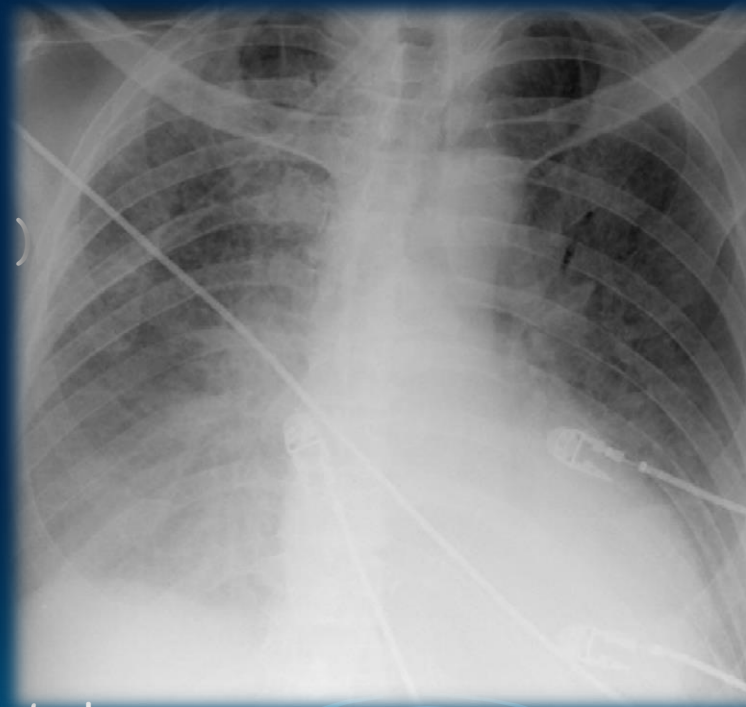
- Dilated LV / Global Hypokinesia

- Severe LV systolic dysfunction

- Ejection Fraction : 26%

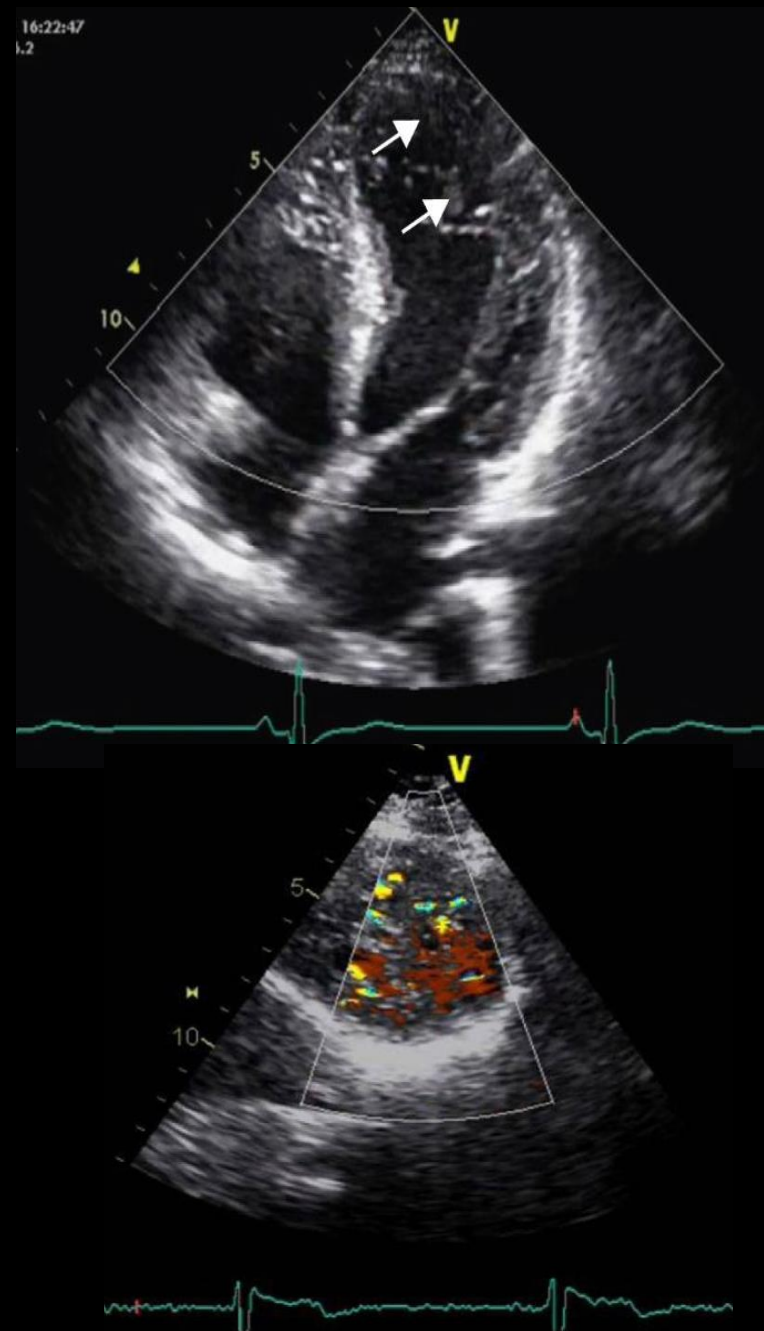
- Mild MR / TR No Thrombus / Embolus

- **? Non Compacted LV**



Apical 4-chamber view,
showing
hyper-trabeculation
of the mid-apical segments
of the lateral wall.

Colour Doppler
showing blood flow
within the
trabecular recesses
of the apex.



Case summary.....

- 8th & 9th Post Operative day
 - Shifted to ward
 - **Repeat 2D Echo – EF of 48%, NCLV**
 - Discharged
 - Tab Ramipril - 5mg/OD
 - Tab Carvedilol - 3.125mg / BD
 - Tab Torsemide 10 mg / OD x 10 days
 - Review with Physician after 10 days
 - Review with Cardiologist after 2 weeks
- Repeat 2DE after 3m - confirmed NCLV

Cardiomyopathies and Pregnancy


- Cardiomyopathies are rare diseases but may cause severe complications in pregnancy
- Etiology of cardiomyopathy in pregnancy -
 - Peripartum Cardiomyopathy (PPCM),
 - Hypertrophic Cardiomyopathy (HCM),
 - Dilated Cardiomyopathy (DCM),
 - Restrictive Cardiomyopathy (RCM),
 - Arrhythmogenic RV Cardiomyopathy (ARVC),
 - Unclassified Cardiomyopathies
 - **LV Non compaction Cardiomyopathy (LVNC)**
 - Alcoholic, Viral, Stress cardiomyopathy
 - Idiopathic cardiomyopathy

Non compacted LV (LVNC)

PREVALENCE

- Very low incidence
- 1 in 2000 echocardiographic studies
- Awareness resulting in increased reporting
- Over / under reporting 2nd diagnostic difficulties

What is Non compaction of LV ?

- NCLV is a rare congenital CMP
 - A loosened spongy myocardium
 - Meshwork of interwoven myocardial bundles
 - Trabeculations with intertrabecular recesses
 - Intra uterine arrest of myocardial compaction
- 

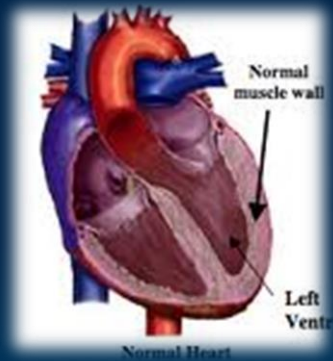
Normal development of LV

- Between 4th to 18th week of gestation
- Majority of myocardium is sponge like
- Endomyocardial trabeculations undergo compaction
- Spongy myocardium Solid

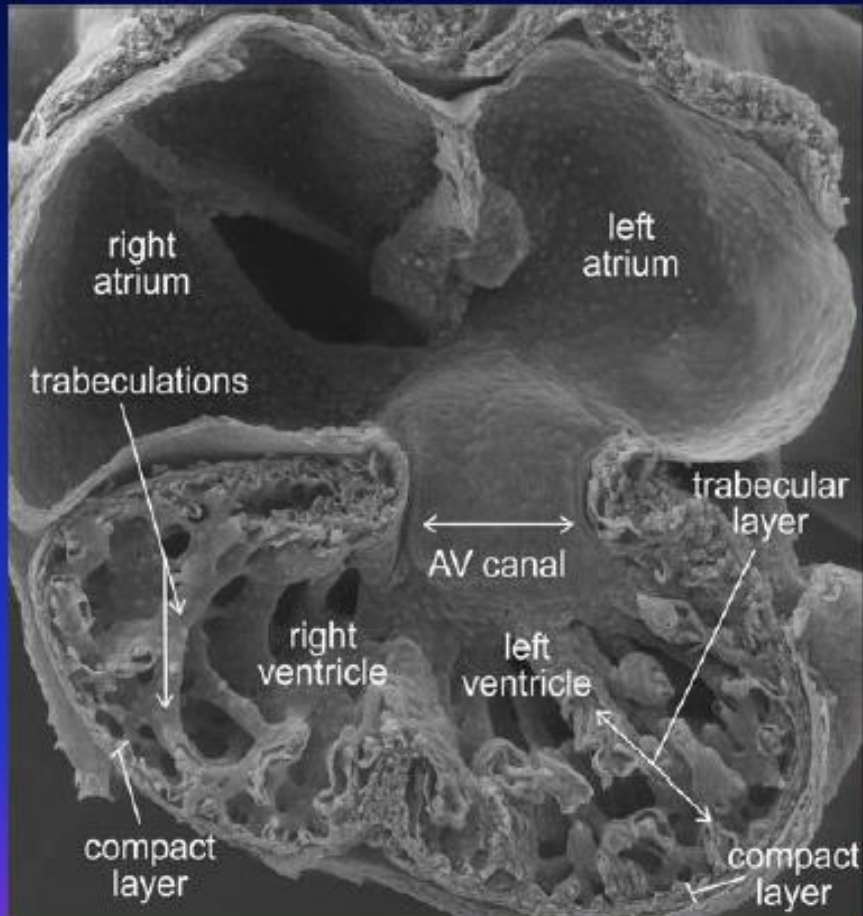


What is Non compaction of LV ?

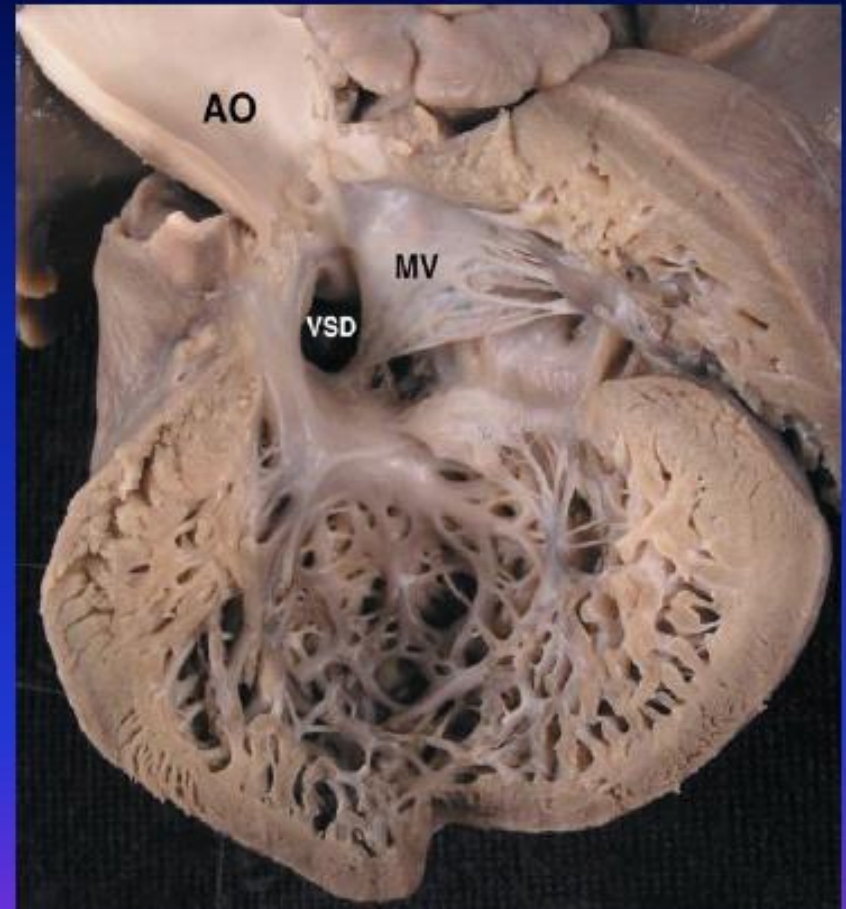
- Normal LV has smooth endocardium and compact myocardium.



Presumed Cause of Non-Compaction

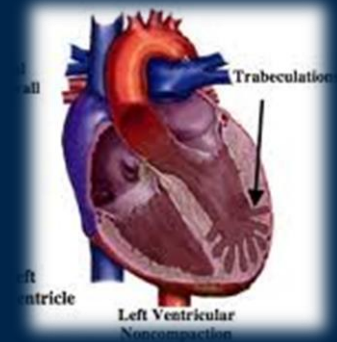


Embryonic Heart

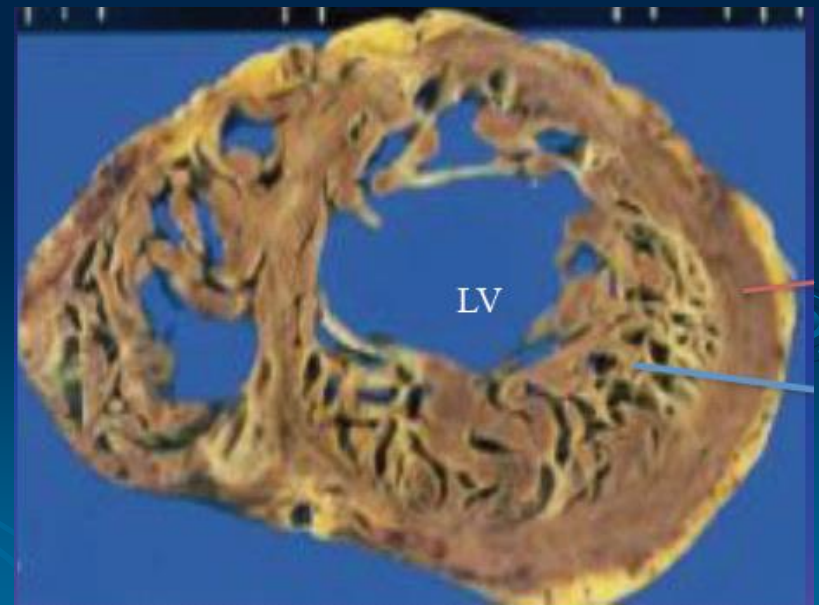


Heart in LVNC

Non compaction of LV



- 2 layered structure of Ventricular wall
- Non compacted Zone
- Compacted Zone
- End systolic ratio > 2



Diagnostic ECHO criteria

- Absence of co-existing cardiac abnormality
- A 2 layered structure of the ventricular wall
- End systolic ratio of non compacted to compacted > 2
- NC in apical and mid ventricular areas
- Direct blood flow from ventricular cavity into the deep inter trabecular recesses (doppler echo)

Non compacted LV (LVNC)

HISTORY

- 1st identified in 1984 - isolated case
- 1st publication by Chin et al in 1980's - 8 cases
- Several publications - single centre case series
- Largest published series -
 - Nation wide survey in France: 2004 - 2006
 - 154 cases reported as LVNC

2004-06 National survey in France

- 154 cases reported as LVNC
- 105 cases reconfirmed as LVNC
- 49 cases the diagnosis was questionable
- Cases followed for 2.33 years
- Common circumstances leading to the Diagnosis
 - CCF in 50%
 - Evaluation of DCM, Rhythm abnormality, Embolic Events
 - Familial recurrence 18-50%
- Age Range: 18 - 86 years
- Male : Female = 66% : 34%
- Prospective Follow-up findings
- Incidence of complications
 - Congestive Cardiac Failure
 - 30% - Recurrent admissions for CCF
 - 4% - Cardiogenic Shock
 - 9% - Cardiac transplant
 - 4% - waiting on transplant list
 - Rhythm abnormalities
 - Embolic Events
 - Heart Transplantation
 - Mortality

Pregnancy & LVNC

- Paucity of cases reported with LVNC & pregnancy (9 so far)
- Physiology in pregnancy
- All suggest -
 - The management of pregnant patients with any inherited cardiomyopathy is directed to the usual treatment of heart failure with diuretics (with or without digoxin) or alternatively hydralazine and nitrates.
 - Early delivery due to heart failure is frequent
 - Preconception evaluation of cardiac function
 - Avoid stressors.
- **References**
 - Pregnancy and treatment outcome in a patient with left ventricular non-compaction
Rahul D. Sawant et al, European Journal of Heart Failure (2013) 15, 592-595
 - Non-compaction cardiomyopathy & pregnancy : An alarming co-existence ending in a favourable outcome
SC plastiras et al, Exp Clin Cardiol, Vol 17, No 3 , 2012

Conclusion

- High index of suspicion to diagnose LVNC
 - Pre-pregnancy assessment
 - Standard heart failure treatment with ACE and aldosterone inhibition together with diuretics and beta-blockers post-delivery is likely to promote recovery of ventricular function once the baby is delivered.
 - Lactation is an additional stressor which may exacerbate heart failure post-delivery
 - Concerted efforts of the team of
 - * Obstetrician
 - * Cardiologist
 - * Anesthetist
 - * Neonatologist
- Mandatory to ensure optimal Outcome