Left Ventricular Noncompaction And Pregnancy – What is New?

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Non-Compaction of Left Ventricle

Cross-section through cardiac ventricles of autopsy specimen.

Ratio of NC/C ≥ 2 is considered diagnostic of LVNC by Echocardiography.
Diagnosis of LVNC

❤ Current imaging studies to diagnose LVNC include:
  ❖ Echocardiography – Most widely used.
  ❖ MRI is being increasingly used to confirm echo.
  ❖ CT scan of the heart
  ❖ Contrast LV Angiography (incidental finding)
  ❖ Video angioscopy. (One case report)

❤ At pathology examination of the explanted hearts show the excessive trabeculations

❤ At autopsy direct examination of the LV.

Diagnostic criteria are not standardized yet.
Echocardiogram in LVNC

Paterick et al JASE April 2012
Diagnosis of LV Noncompaction

Videoangioscopy

MRI of heart with LVNC
Contrast Angiogram of LV in Noncompaction

Arrows pointing to the deep trabeculations and contrast filling between them.
What are the complications in LVNC?

Major complications are:

❤️ **Heart failure:**
- Both systolic and diastolic HF can occur.

❤️ **Arrhythmias:**
- Patients may be on anti-arrhythmic medicines, have pacemakers, ICDs etc., or may have had RF ablation for atrial or ventricular arrhythmias.

❤️ **Thromboembolism:**
- Both systemic and pulmonary embolism can occur.
25 reports from 2003 to 2014: 33 women
Personal Communications: 2 patients

54 Pregnancies in 35 women: (Ages 14 to 41 years).
Multiple pregnancies in LVNC patients:
  2 pregnancies  8 patients
  3 pregnancies  4 patients
  4 pregnancies  1 patient

One maternal death with cerebral infarct after C-section, reported from China Papers (Master’s thesis).

Fetal/neonatal mortality:
  1 intrauterine death at 29 weeks.
  1 neonate died 2 days post C-section at 24 weeks.
  2 Abortions (at 5 and 11 weeks).
<table>
<thead>
<tr>
<th>Procedure</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Delivery by C-Section</td>
<td>18</td>
</tr>
<tr>
<td>Delivery 2 times C section</td>
<td>3</td>
</tr>
<tr>
<td>Heart Transplant</td>
<td>2</td>
</tr>
<tr>
<td>L.V. Assist Device</td>
<td>2</td>
</tr>
<tr>
<td>IABP</td>
<td>2</td>
</tr>
<tr>
<td>ICD implanted</td>
<td>5</td>
</tr>
<tr>
<td>R.F ablation (Atr.flutt &amp; VT)</td>
<td>1</td>
</tr>
<tr>
<td>ECMO stand by</td>
<td>1</td>
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</tbody>
</table>

1 other patient had an ICD 33 months prior to pregnancy;
Medical Management of Heart Failure

- **Beta blockers**: Metoprolol, bisoprolol, carvedilol
- **ACE Inhibitors**: Enalapril, Lisinopril, Candesartan
- **Diuretics**: furosemide, spironolactone, amiloride, Nesiritide.
- **Inotropes**: Digoxin, Milrinone, Dobutamine
- **Anticoagulation**: Warfarin, LMW Heparin
- **Aspirin**
- **Bromocriptine.** (Inhibits prolactin secretion)
- **Antiarrhythmic drugs**: Procainamide, Amiodarone.
LVNC and Peripartum Cardiomyopathy – 11 cases Reported

❤ Bahl, Lea, Rehfeldt and Peters each described a case of LVNC with heart failure, who also met criteria for PPCM as well as LVNC; (4 cases)

❤ Patel et al described 2 cases of LVNC with heart failure initially diagnosed as peripartum cardiomyopathy.

❤ Rajagopalan reported a series of 5 cases that met criteria for diagnosis of PPCM as well as LVNC.

❤ It appears that peripartum cardiomyopathy has worse prognosis compared to other dilated cardiomyopathies; hence it is important to distinguish these conditions.

Management of Complications

❤ Echocardiographic assessment is useful to guide therapy in complex cases. Biomarkers may also be helpful.

❤ No specific recommendations for LVNC; drug treatment is same as for other dilated cardiomyopathies in pregnant women.

❤ Digitalis, beta blockers like carvedilol or metoprolol are safe (avoid atenolol).

❤ Diuretics are safe if they do not cause dehydration & placental hypoperfusion and oligohydramnios.

❤ Antiarrhythmic drugs & devices as needed.

ESC Guidelines on Management of Cardiovascular disease during pregnancy 2011
Sarma RJ, Progress in Cardiovascular Diseases. 52 (4) 264-273, 2010.
Krul, European Journal of Heart Failure (2011) 13, 584–594
Anesthetic Considerations for C-section

❤ General or epidural anesthesia is used commonly.
❤ Epidural anesthesia offers some advantages:
  ❖ Easy conversion from labor analgesia to surgical anesthesia.
  ❖ Preservation of fetal and maternal hemodynamics.
  ❖ Prevention of increase in catecholamines.
  ❖ Possible suppression of arrhythmias due to pharmacologically-active plasma levels of local anesthetic.
❤ Turning the ICD off or on did not seem to affect the outcome in one study. If electrocautery is to be used ICDs are turned off.
❤ ECMO standby for decompensated HF patients.
Summary

- LVNC has not yet been identified as a distinct cardiomyopathy by the (WHO) World Health Organization or (ESC) European Society of Cardiology.

- Diagnostic criteria are not standardized yet.

- Pregnancy is not uncommon in patients with isolated LVNC. More experience is being published on this topic.

- Peripartum cardiomyopathy may occur in patients with LVNC.

- Management of complications is not specific to LVNC.

- Family screening using echocardiography is important to detect this genetically mediated cardiomyopathy in the newborn as well as other family members.
What Else is new with LVNC in 2014?
Registries?
The Social media?
Support Groups?
Many Registries with data on Noncompaction

- German NCCM Registry (ALKK)
- German - Austrian Left Ventricular Noncompaction Registry (2007)
- French Registry – 2004 to 2006. (105 cases included)
- S I E C (Societa' Italiana di Ecografia Cardiovascolare). 2004 -
- Australian Genetic Heart Disease Registry....
HeduAfrica on Facebook.
Left Ventricular Noncompaction (LVNC)

What is left ventricular noncompaction (LVNC)? PDF version

Left ventricular noncompaction (also known as LVNC) is a disease of the heart muscle that has only recently been described. Left ventricular noncompaction can occur on its own (i.e. isolated) or along with other heart problems (i.e. congenital heart disease) and is characterised by deep trabeculations (finger-like projections) in the muscle wall of the left ventricle. These trabeculations can also occur in the right ventricle. The heart muscle abnormalities occur during the development of the heart in the embryo. Symptoms of the disease are variable; with some patients having no symptoms while others may develop shortness of breath, palpitations, chest pain, dizziness and fainting episodes. Occasionally the disease can cause heart failure, stroke (due to blood clots forming in the trabeculations then travelling to the brain) or sudden death.

Prof Chris Semsarian
Chair, Registry Advisory Committee
**Left ventricular noncompaction**

Left ventricular noncompaction cardiomyopathy is a heart muscle condition in which the myocardium is not "compacted", consisting of a meshwork of numerous muscle bands called trabeculae cardiomyopathy.

This type of cardiomyopathy has not been fully understood so far and remains under development, clinical course and treatment are fields of ongoing research.

**The development of the heart**

The foetal heart muscle has a non-compacted appearance between the 4th and 5th month of life when the heart is nourished by the 4 coronary arteries, which will eventually take over carrying blood to the heart.

The thickness of the compacted wall and the mass of the heart muscle is greater than in the compacted muscular wall of the heart with minor muscle bands close to its apex.

Some limited data suggests that similar cardiac features can be acquired with other congenital cardiac malformations. Experimental studies have shown problems with the heart muscle function with impaired pumping strength, and some of these defects can lead to heart failure. More recent studies, though, have demonstrated wider differences in severity and outcome in patients with heart failure.

**Prevalence and diagnosis**

The number of people with this condition is thought to be very low and it has been found in one in 2,000 echocardiograms. It is evaluated and diagnosed by most of the researchers and clinicians, is by quantifying the size of the muscle.

The ECG may reveal abnormalities but these are usually non-specific, whereas imaging of the heart, such as echocardiography, shows that the diagnosis of the disorder may be neither easy nor objective in many cases and has recently been a topic of significance with other heart muscle disorders.
Welcome to the Non Compaction Cardiomyopathy Association! The purpose of this site is to share information and to help connect patients (and families of patients) that have been diagnosed with Non Compaction.

Jeffrey A. Towbin, MD Researcher at Cincinnati Children’s Hospital
LVNC is on FACE BOOK now. 312 members

NCC Assoc. has two Facebook locations.
- One public page for info and Association related info.
- One private page for patient conversations and support.

If you need any assistance or have questions about these pages, please contact noncompaction@gmail.com

The NCC Association also has a page on Facebook. It is a great place to share links and info. It also serves as a public page to raise awareness and share with others who want to show support but may not be interested in the conversations on the private page.

www.facebook.com/noncompaction
The Children’s Cardiomyopathy Foundation works in partnership with the National Heart, Lung and Blood Institute funded North America Pediatric Cardiomyopathy Registry (PCMR). This patient registry was established to provide a population base to describe the epidemiologic features and clinical course of selected cardiomyopathies in patients aged 18 years or younger, and to promote the development of etiology specific treatments.

The PCMR, established in 1994, is a confidential database of children diagnosed with cardiomyopathy and is the only national registry of its type in North America. The database consists of 3,000 enrolled patients from 100 medical centers in the United States, Canada and Puerto Rico. The University of Miami is the PCMR administrative coordinating center and the New England Research Institute serves as the PCMR’s data coordinating center.
Similarity to HCM

Source Paterick et al: JASE April 2012

(DOI:10.1016/j.echo.2011.12.023)

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# LVNC: The MVP of the 21st Century?

Gabriella Captur, Andrew S. Flett, Daniel L. Jacoby, James C. Moon

## Table 2: Comparing MVP and LVNC

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<tr>
<th></th>
<th>MVP</th>
<th>LVNC</th>
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</table>
| **Subtypes**         | Classic versus nonclassic  
|                      | Symmetric versus asymmetric  
|                      | Flail versus non-flail                                                | None  
|                      |                                                                         | No ethnic-specific reference ranges |
| **Diagnostic criteria** | 2/3D Echo and TOE  
|                      | ≥2mm billowing  
|                      | ≥5mm thickening                                                      | 2D Echo (5 techniques)  
|                      | Established                                                        | CMR (2 techniques)  
|                      |                                                                         | Consensus lacking |
| **Incidence**        | Historically  
|                      | M-mode 17%  
|                      | 2D Echo 35%  
|                      | Presently  
|                      | 2-3%                                                      | True incidence unknown |
| **Associations**      | Myxomatous degeneration  
|                      | Marfan’s syndrome  
|                      | Ehlers-Danlos syndrome  
|                      | Osteogenesis imperfecta  
|                      | Polycystic kidney disease                                           | Genetic  
|                      |                                                                         | Neuromuscular disorders  
|                      |                                                                         | Mitochondrial myopathies  
|                      |                                                                         | Barth syndrome  
|                      |                                                                         | Zaspopathy  
|                      | Well established                                                    | Cardiac  
|                      |                                                                         | Dilated cardiomyopathy  
|                      |                                                                         | Hypertrophic cardiomyopathy  
|                      |                                                                         | Restrictive cardiomyopathy  
|                      |                                                                         | Congenital heart disease  
|                      |                                                                         | Conduction disease  
| **Complications**     | Severe mitral regurgitation  
|                      | Atrial fibrillation  
|                      | Heart failure  
|                      | Ischaemic neurological events  
|                      | Infective endocarditis  
|                      | Mitral prolapse syndrome  
|                      |                                                                         | Arrhythmias  
|                      |                                                                         | Heart failure  
|                      |                                                                         | Thromboembolic events  
|                      |                                                                         | Sudden cardiac death  
|                      |                                                                         | Reported frequencies  
|                      |                                                                         | vary (based on single-centre case series) |
| **Prognostic determinants** | Specific to MVP  
|                      | Chordal rupture/flail leaflet  
|                      | Related to associated pathology  
|                      | Severity of MR and ERO  
|                      | LV systolic dysfunction                                                | Specific to LVNC  
|                      |                                                                         | None known  
|                      |                                                                         | Related to associated pathology  
|                      |                                                                         | LV systolic dysfunction  
|                      |                                                                         | Atrial fibrillation  
|                      |                                                                         | Congestive heart failure  
|                      |                                                                         | Coexistent neuromuscular disorders  
|                      |                                                                         | Data from one single-centre case series  
|                      |                                                                         | Single-centre: largest n = 1628  
|                      |                                                                         | Multi-centre registry: largest n = 105  
| **Study methodology** | Case reports  
|                      | Case series  
|                      | Population-based case-control studies  
|                      | Randomised control studies                                            | Case reports  
|                      | Case series  
| **Treatment**        | Sequential refinements in mitral valve repair procedures  
|                      | Mitral valve replacement                                              | Consensus lacking  

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The above table summarizes the similarities and differences between MVP and LVNC, considering various aspects such as subtypes, diagnostic criteria, incidence, associations, complications, prognostic determinants, study methodology, and treatment.
What needs to be done now?

“The establishment of an International Registry would help bring together these disparate sources of information, with a view to producing universally accepted guidelines for the diagnosis and treatment of these patients”.

Quoted by:
What needs to be done now?

Prospective studies are needed to document the natural progression of LVNC and to determine clinical and imaging predictors of adverse outcomes, which could be the basis for a simple diagnostic prediction rule.

This is a task for a coordinated effort between the international imaging and heart muscle disease communities to come together to plan, raise funds for, and organize such a study.

By Steffen E Petersen, MD, DPhil.
London, United Kingdom
Thank you for your attention.