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Left Ventricular Noncompaction: Case report

Dinesh Bisht[1], Neeraj Awasthy[2]
[1, 2] Pediatric Cardiologist, Pediatric Cardiology Dependent, Max Hospital Saket, Delhi, India

Abstract
Left Ventricular Non-Compaction (LVNC) is a rare congenital cardiomyopathy that is characterised by thin compacted epicardial layer and extensive non-compacted endocardial layer with prominent trabeculations and deep recesses. It may vary from mild to severe forms and may present at any age. It has a strong hereditary and genetic predisposition and variable penetrance. It is manifested as CHF, arrhythmia or DCM. It can be diagnosed by Echocardiography, although MRI is superior modality. The treatment includes beta-blockers, antiarrythmics, decongestive drugs, ICD and anticoagulants. Most of LVNC patients (severe forms) need heart transplantation.

Keywords: ventricular, cardiomyopathy, noncompaction, congenital

Introduction: Case Report

We report a family of 10 sibs; 8 of them have various degrees of LVNC. Six were females; out of six females two were normal; other four had mild apical LVNC. Youngest kid is 2-year-old and eldest is 22-year-old. Out of four males, one died at 15 years of age, he was suffering from dilated cardiomyopathy with CHF due to severe LVNC. Second male sib has LVNC with LVEF=40% and LV is dilated. Third male sib is healthy. The fourth male sib, the youngest one is two years old also has mild apical LVNC.

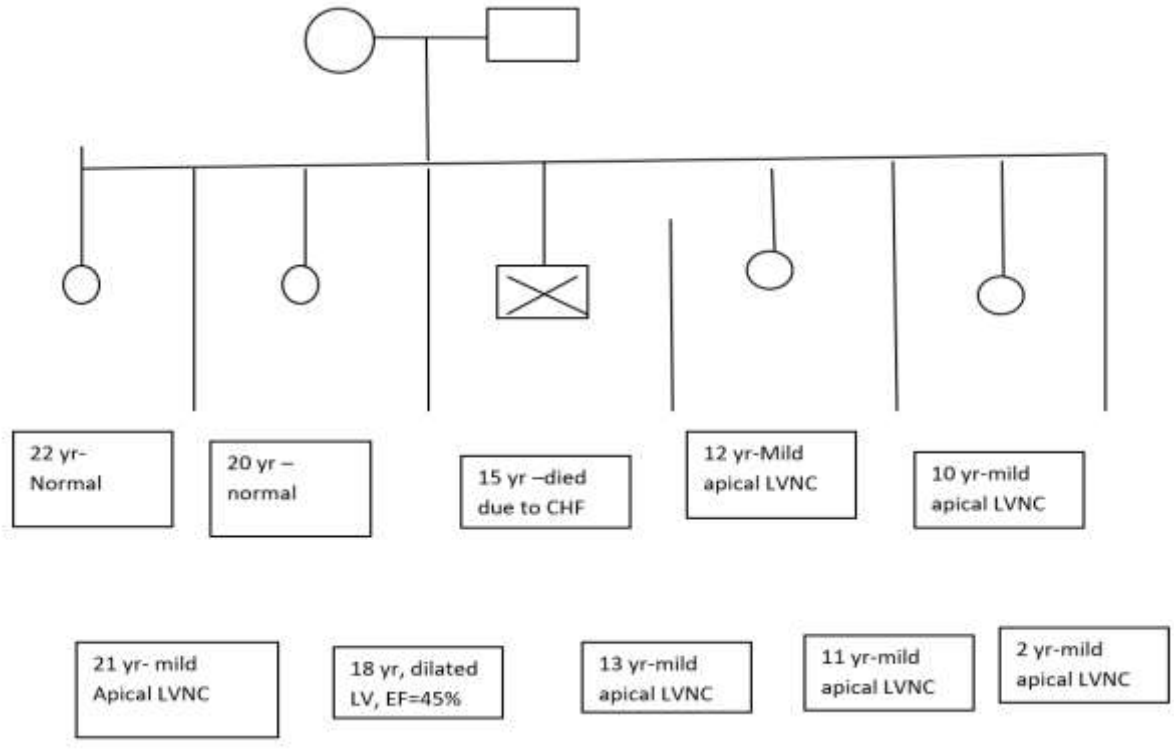

Fig 1

Conclusion

LVNC has never been reported in the past in such a large family of 10 children. This suggests that LVNC has immense hereditary predilection and is expressed in male and female phenotypes; although in males the presentation is worse as we know X-linked mutant gene is the culprit. However, LVNC can present in various forms of severity varying from mild apical LVNC to severe DCM resulting in death. In the same family one male and one female sib is completely healthy, thus concluding that genetic penetration of LVNC is not 100%.

Discussion

LVNC or spongy myocardium is a rare congenital cardiomyopathy that can be diagnosed at any age. It is characterised by a thin, compacted endocardial layer with

prominent trabeculations and deep recesses that communicate with LV cavity but not with coronary circulation, probably due to an arrest of compaction during intra-uterine life. The prevalence is 0.014- 1.3% in general population. Eventually this condition can potentially lead to chronic heart failure, arrhythmia and embolic events. Echo diagnostic criteria: - End systolic ratio of non-compacted to compacted myocardium layers >2. MRI criteria: -1. Noncompacted to compacted myocardium ratio>2.3. 2. Trabeculated LV mass>20% of total mass. Amount of delayed trabecular enhancement correlates significantly with LVEF. RV noncompaction has also been reported in less than half patients. Endomyocardial biopsy shows interstitial fibrosis in all necrotic myocytes in prominent trabeculations, endomyocardial thickening and subendocardial fibroelastosis has also been found. Fetal echo was normal in LVNC.

Familial occurrence is seen in 18%. Mutation in G4.5 X-linked gene has been identified. Subendocardial perfusion defects have been described using MRI. PET and Thallium scintigraphy have demonstrated transmural perfusion defects correlating with areas of noncompacted myocardium. Post-mortem studies have discovered ischemic sub-endocardial lesions; thus, supports the hypothesis that coronary microcirculation abnormalitie may play a key role in its pathophysiology leading to contractile dysfunction. Diastolic dysfunction may be related to abnormal relaxation and restrictive filling caused by numerous trabeculea. High percentage of associated neuromuscular disorders have been described in patients with LVNC. 60% patients die within 4 years of followup and 90% develop LV dysfunction within a period of 10 years of followup.

References

1. Bennett CE, Freudenberger R. The Current Approach to Diagnosis and Management of Left Ventricular Noncompaction Cardiomyopathy: Review of the Literature. Cardiol Res Pract, 2016. 2016:5172308. doi: 10.1155/2016/5172308. Epub 2016 Jan 11. Review.
2. Dong X, Fan P, Tian T, Yang Y, Xiao Y, Yang K, *et al*. Recent advancements in the molecular genetics of left ventricular noncompaction cardiomyopathy. Clin Chim Acta. 2017; 465:40-44. doi: 10.1016/j.cca.2016.12.013. Epub 2016 Dec 15. Review.
3. Finsterer J, Stöllberger C, Towbin JA. Left ventricular noncompaction cardiomyopathy: cardiac, neuromuscular, and genetic factors. Nat Rev Cardiol. 2017; 14(4):224-237. doi: 10.1038/nrcardio.2016.207. Epub 2017 Jan 12. Review.
4. Towbin JA, Lorts A, Jefferies JL. Left ventricular non-compaction cardiomyopathy. Lancet. 2015; 386(9995):813-25. doi: 10.1016/S0140-6736(14)61282-4. Epub 2015 Apr 9. Review.
5. Wang C, Takasaki A, Watanabe Ozawa S, Nakaoka H, Okabe M, Miyao N, *et al*. Long-Term Prognosis of Patients with Left Ventricular Noncompaction - Comparison Between Infantile and Juvenile Types. Circ J. 2017; 81(5):694-700. doi: 10.1253/circj. CJ-16-1114. Epub 2017