CASE REPORT

A RARE CASE OF FETAL CARDIAC SYNDROME

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HOW TO CITE THIS ARTICLE:
G. Arun Kumar, A. R. Malathy, Jemima Bhaskar, Nalini V. "A Rare Case of Fetal Cardiac Syndrome". Journal of Evidence Based Medicine and Healthcare; Volume 1, Issue 7, September 2014; Page: 782-786.

ABSTRACT: A 42 year old male patient, who is not a smoker presented with chief complaints of breathlessness on mild exertion, chest pain for past 2 months. Initially it was suspected to be coronary artery disease which presented as cardiac failure. We admitted the patient and started on diuretics and anti-failure measures. After stabilization, echocardiogram was done. It showed non compaction of left ventricle**.** Non-compaction of the ventricular myocardium (NVM) is an uncommon disorder. It is thought to be caused by arrest of the normal process of endo myocardial morphogenesis. Jennes diagnostic criteria were fulfilled. Hence the diagnosis of idiopathic non compaction of left ventricle (INVM) was confirmed. Detailed assessment of cases presenting with cardiac failure helps us to arrive at such unusual diagnosis. The screening of family members prevents sudden death.

KEYWORDS: Non-compaction of left ventricle, Jennes diagnostic criteria.

INTRODUCTION: IDIOPATHIC LEFT VENTRICULAR MYOCARDIUM is often missed as a diagnosis. It is a formidable challenge to diagnose this condition and is often under evaluated for its etiology. Hence high index of suspicion is a must in all such cases.

CASE REPORT: A 42 year old man presented with chief complaints of dyspnoea Grade II with palpitation on & off. He gives no history of chest pain, syncopal attacks fever, wheeze and cough with expectoration.

Past history: not a known case of DM/SHT/CAD
Personal history: not a smoker, occasional alcoholic, bowel bladder habits –N
Family history:

No h/o sudden death in the family.

On examination patient conscious, oriented, dyspnoeic at rest, no pallor, no cyanosis, mild pedal edema.

Vitals: BP-110/70, PR-114/mt, regular RR-20/mt

CVS-S1S2+S3 gallop+, no murmurs
RS-NVBS
P/A-soft, no free fluid
No organomegaly
CNS-NFND

INVESTIGATIONS:

Routine investigations –normal

ECG-NSR, Left ventricular hypertrophy (by voltage criteria), symmetric

t wave inversion in all leads

Chest x ray- cardio thoracic ratio 55%

cardiomegaly +

USG abdomen –normal study

2D ECHOCARDIOGRAM –ISOLATED NON COMPACTION OF LEFT VENTRICLE, EF-42%, mild LV diastolic dysfunction.

Treatment:

- Salt restricted diet
- Fluid restriction
- Diuretics
- Beta blocker
- Ace inhibitor

Patient improved symptomatically.

Patient's parents, children and siblings were screened and found to be normal.

DISCUSSION: Non compaction of the ventricular myocardium is a cardiomyopathy thought to be caused by arrest of normal embryogenesis of the endocardium and myocardium. This abnormality is often associated with other congenital cardiac defects, but it is also seen in the absence of other cardiac anomalies. Echocardiography has been the diagnostic procedure of choice, but the correct diagnosis is often missed or delayed because of lack of knowledge of this condition.

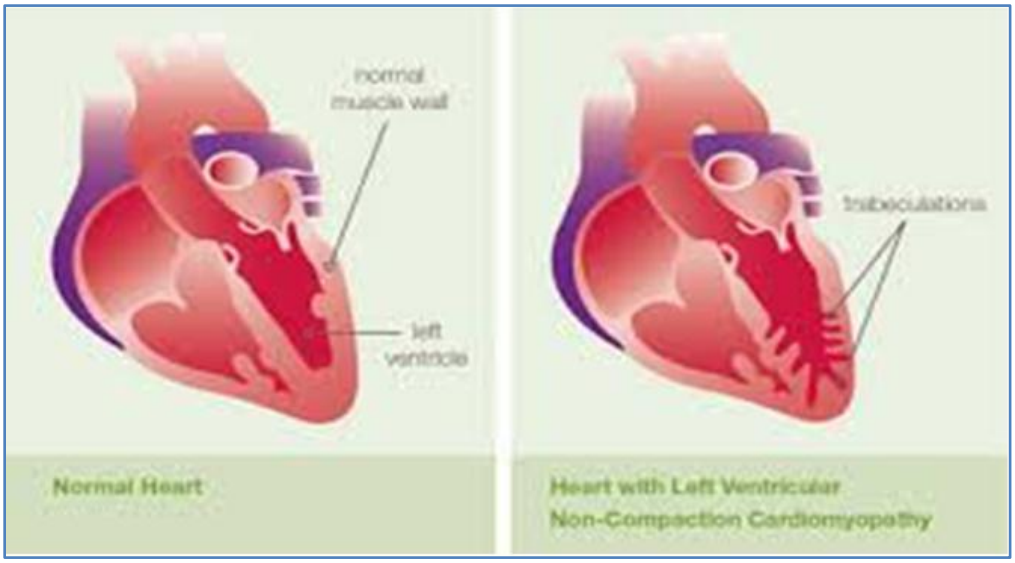

Fig. 1.a: Normal heart figure1.b.Heart with INLV

EMBRYOLOGY: Gradual "compaction" of spongy meshwork of fibers and inter trabecular recesses, or "sinusoids," occurs between weeks 5 and 8 of embryonic life, proceeding from the epicardium to endocardium and from the base of the heart to the apex.[1–5] The coronary circulation develops concurrently during this process, and the inter trabecular recesses are reduced to capillaries. The normal process of trabeculation appears to involve secretion of neuregulin growth factors.[6]

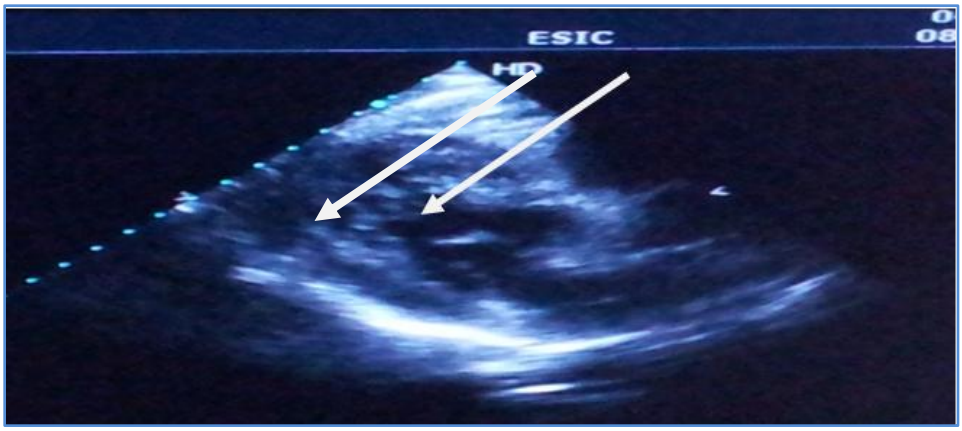

Fig. 2: 2D Echocardiogram shows sinuses in the left ventricle

Jennes diagnostic criteria: A quantitative evaluation for the diagnosis of INVM by determining the ratio of maximal thickness of the non-compacted to compacted layers (measured at end systole in a parasternal short axis view), with a ratio >2 diagnostic of INVM.

Both familial and sporadic forms of non-compaction have been described. In the original report of INVM, which predominantly involved children, familial recurrence was seen in half of patients. Mutations in LDB3 (cypher/zasp) gene, 1q21.1 deletion syndromes are also identified.

The median age at diagnosis was 7 years (ranging from 11 months to 22 years). In the largest series of patients with INVM,[8] the prevalence was 0.014% of patients referred to the echocardiography laboratory. It may present in elderly Men appear to be affected more often than women, with males accounting for 56% to 82% of cases.[1, 3, 8, 23]

Pathophysiology: Diastolic dysfunction in ventricular non-compaction may be related to both abnormal relaxation and restrictive filling caused by the numerous prominent trabeculae.[2] The origin of systolic dysfunction in non-compaction is unclear, but a body of evidence is accumulating that point toward sub endocardial hypo perfusion and microcirculatory dysfunction playing roles in ventricular dysfunction and arrhythmogenesis. Because of the prominent, numerous trabeculae, subendocardial ischemia may result from isometric contraction of the endocardium and myocardium within the deep intertrabecular recesses.

Complications: Arrhythmias. Atrial fibrillation has been reported in over 25% of adults with this IVNM. Ventricular tachyarrhythmias have been reported in as 47%.Sudden cardiac death accounted for half of the deaths in the larger series of patients with INVM.[1, 3, 8, 9] Abnormalities of

the resting ECG are found in the majority of patients with NVM but findings are nonspecific and include left ventricular hypertrophy, repolarization changes, inverted T waves, ST segment changes, axis shifts, intraventricular conduction abnormalities, and AV block.[1] Isolated NVM, the occurrence of thromboembolic events, including cerebrovascular accidents, transient ischemic attacks, pulmonary embolism, and mesenteric infarction, ranged from 21% to 38%.Embolic complications may be related to development of thrombi in the extensively trabeculated ventricle, depressed systolic function, or the development of atrial fibrillation.[1] Of interest, no systemic embolic events were reported in the largest pediatric series with INVM.[2]

CONCLUSION: Any patient presenting with cardiac failure in any age and history of sudden death in family members, non-compaction of ventricular myocardium should be suspected. Earlier the diagnosis and screening of family members may prevent the complication and sudden death in the family. IVNM usually presents in the pediatric population.

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Date of Submission: 16/08/2014.
Date of Peer Review: 18/08/2014.
Date of Acceptance: 21/08/2014.
Date of Publishing: 16/09/2014.