myocardium in association with a left ventricular aneurysm. Pericardi- 
cardial calcification was found primarily over the right-sided car-
diac chambers and in the atrioventricular grooves, infrequently 
on the apex of the left ventricle. When the left ventricle was involved, there was always 
more extensive calcification elsewhere in the pericardium. Myocar-
dial calcification occurred predominantly in the apex of the left 
ventricle, although it was rarely confined to the posterior wall 
of the left ventricle. Isolated calcification in the region of the left ven-
tricular apex, therefore, strongly suggests left ventricular aneurysm. We present a 45 year old patient with old anterior wall myocardial 
infarction who had significant left ventricle wall calcification with 
no aneurysm. No calcium or parathyroid abnormalities could be 
detected in the patient after lab investigations. Metastatic calcification of various organs including myocardium 
has been reported with HTLV-1 infection, but in our case, HTLV-1 
was negative. Myocardial calcification has been reported after 
orthotopic heart transplantations or unselected bone marrow 
transplantation to acute myocardial infarction in an animal model. 
Thus isolated idiopathic left ventricular calcification without any 
detectable abnormality was found to be worth reporting.

A case of Marfans syndrome with 
ascending and arch of aorta aneurysm 
presenting with type A dissection 
of aorta

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Introduction: Marfans syndrome is a hereditary disease which is 
autosomal dominant inheritance because of mutation in the fibril-
lin-1 gene, which affects connective tissue of the body, mainly 
involves cardiovascular system, ocular and skeletal system. It is 
usually diagnosed with 2010 Revised Ghent Nomenclature with a score 
of more than 7. The cardiovascular manifestation includes aortic root 
and arch aneurysm with high risk of dissection (root diameter of 
>4.5 cm), mitral valve prolapse, aortic regurgitation secondary to 
root dilatation, skeletal abnormalities scoliosis, pectus excavatum, 
positive thumb & wrist sign, ectopia lens. 
Case report: A female 40 years old patient presented with chest pain 
retrosternal since 1 month, tearing type radiating neck and back of 
chest associated with breathlessness of class 3 on examination 
conscious and coherent pulse rate = 78/min felt in all limbs, blood 
pressure 150/60 mm of Hg, CVS S1 S2+, ESM 3/6 and EDM heard on 3rd 
left intercostal area, MDM at the apex. ECG was showing LV volume 
overload, on 2D echo evaluation was showing aorta root showing 
4.78 cm with severe AR good LV function. Patient was evaluated with 
trans-esophageal echo, chest X ray and CECT was diagnosed as 
ascending and arch aneurysm with dissection of aorta type-A. 
Patient was referred to CT surgery dept for Benthal procedure.

Discussion: The incidence of aortic dissection is estimated to be 2– 
3.5/10,000 persons per year and peak incidence at sixth and 
seventh decade with overall mortality 1%/h, patients with Marfans 
syndrome at higher risk can occur at younger age. It is classified 
into Debakey type 1,2,3 and Stanford type A and B depending 
upon location of dissection. High clinical suspicion required for 
diagnosing dissection has variable clinical manifestation most 
common is chest pain (80%), severe aortic insufficiency (45%), 
hypotension (14%), shock (13%), syncope (12%), MI (7–19%), CVA 
(8%) and paraplegia (2%), pulse deficit is seen (26%). The manage-
ment of the dissection is beta blocker drug of choice, followed by 
ACE inhibitors. Medical management is considered in uncompli-
cated and chronic type B dissection, surgery is the treatment of 
choice acute type A, complicated type B, associated with Marfans 
syndrome, end organ dysfunction. Endovascular therapy can be 
done in alternative in complicated type B dissections. 
Conclusion: Reporting a case Marfans syndrome with type A dis-
section diagnosed TEE.

Ventricular dysynchrony with VVI pacemakers – Does complete heart 
block patients differs from sinus node 
dysfunction?

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Background of the study: To assess the ventricular dysynchrony in VVI pacemaker patients and to find any difference in occurrence of 
ventricular dysynchrony between sick sinus syndrome and complete heart block patients. 
Materials: Totally 16 patients who underwent VVI pacemaker were studied. The indications for permanent pacemaker implantation 
were complete heart block and sick sinus syndrome. Among 18 
patients, 14 patients had diagnosis of complete heart block and 4 
patients had sick sinus syndrome. The follow up assessment of 
ventricular dysynchrony was done with a minimum period of one 
month and maximum period of 12 years after the VVI pacemaker 
implantation. Patients who had myocardial infarction, valvular 
heart disease, cardiomyopathies and regional wall motion 
abnormalities at the baseline were excluded from the study. 
Methods: All the 18 patients underwent 2D echo, Doppler, M-mode 
in the HD 7 Phillips echo machine for assessing the ventricular 
dysynchrony, intraventricular dysynchrony, interventricular dysynchrony and atrioventricular dysynchrony were assessed. 
Intraventricular dysynchrony was defined if the septal to posterior 
wall delay is more than 130 ms. Interventricular dysynchrony 
was diagnosed if the difference in pre-ejection time interval between 
two ventricles was more than 40ms. Atrioventricular dysynchrony 
was diagnosed if the ratio of left ventricular filling time to RR interval is less than 40%. 
Results: After the study, 6 patients with complete heart block had 
significant intraventricular dysynchrony with septal to posterior wall 
delay of more than 130 ms. By comparing the results between com-
plete heart block and sinus node dysfunction patients, none of the 
n sinus node dysfunction patients developed intraventricular dyssyn-
chrony. The intraventricular dysynchrony is much prolonged, with 
the value of more than 155 ms in patients who underwent the VVI 
pacemaker 12 years back. None of the patients who underwent the 
study had interventricular and atrioventricular dysynchrony. 
Conclusion: Following VVI pacemaker intraventricular dyssyn-
chrony is common. But this study reveals intraventricular dyssy-
nchrony is more common in complete heart block than sinus node 
dysfunction patients. The reason for intraventricular synchrony in 
sinus node dysfunction is not clear, even though both groups were 
paced from right ventricle.

Echocardiographic assessment of LV clot and its DDs

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Background: Cerebrovascular accident (CVA) is one of the leading 
cause of mortality and morbidity across the globe irrespective of
component of LV myocardial deformation in hypertension and therefore most sensitive to the presence of myocardial disease. In hypertensive patients, abnormalities in baseline myocardial deformation are identified in patients with altered left ventricular geometry. Myocardial strain varies depending on the left ventricle’s degree of remodeling and systolic function. Myocardial deformation indices may play a role in reflecting the mechanisms linking altered left ventricular geometry with progression to decompensated left ventricular systolic function.

Cardiac amyloidosis, Congo Red negative: Diagnostic error or a disease begging to be diagnosed?

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Introduction: Restrictive Cardiomyopathy (RCM) is a well known entity characterized by progressive diastolic and subsequently systolic biventricular dysfunction. Of the specific causes of RCM, infiltrative disorders are easily recognizable and have specific characteristics. Amyloidosis tops the list among infiltrative disorders presenting as RCM. We report a series of 7 patients with amyloidotic and non amyloidotic infiltrative cardiomyopathies, their presentation, diagnosis and implications in the management.

Case series report: Our series includes 7 patients (5 patients were males and 2 were females, with age of 46–58 years). There was no history of hypertension or diabetes. Only one patient had known multiple myeloma and presented subsequently with heart failure and RCM – AL Amyloidosis. 4 patients were diagnosed elsewhere to have non ischemic dilated cardiomyopathy and 2 other patients were diagnosed with hypertrophic cardiomyopathy. On evaluation, they were diagnosed to be having RCM with possible infiltrative etiology. All had low voltage ECG complexes despite severe Biventricular hypertrophy on the echocardiogram. Echocardiography revealed biventricular hypertrophy, speckled hyperechocoid myocardium, thickened inter atrial septum, restrictive pattern of diastolic dysfunction, varying degrees of mitral and tricuspid regurgitation and pulmonary arterial hypertension. Four of them also had severe LV systolic dysfunction with EF of 20–35%. Three patients had reduced global longitudinal strain with “apical sparing” and relative preservation of radial and circumferential strain, a feature in favor of amyloidosis. Free light chains (kappa or lambda) were elevated in all. Serum immunoelectrophoresis revealed monoclonal gammapathy in four. Urine Bence Jones protein was elevated in two. All but one had Congo Red positivity on bone marrow, the remaining patient was Congo Red negative even on endomyocardial biopsy. Non amyloidotic light chain deposition disease was diagnosed by exclusion (reports of electron microscopy and immunohistochemistry are awaited). 6 of them had multisystem involvement (renal dysfunction and proteinuria, hepatic dysfunction).

Five were initiated on chemotherapy with advice for heart transplantation together with Bone marrow transplantation and chemotherapy (Bortezomib regimens). Four died in 6 months from the time of presentation and 3 are on follow up with results of electron microscopy and immunohistochemistry awaited in one.

Conclusion: Cardiac amyloidosis is an underappreciated entity. After our diagnosis of first case of non amyloidotic light chain deposition disease, we have been more systematic in analysing patients diagnosed with RCM. Establishing an etiological diagnosis is very important for initiating appropriate therapy including specific pharmacological measures & has an impact on organ transplantation advice and prognostication.