SNS COLLEGE OF ALLIED HEALTH SCIENCES- COIMBATORE 35

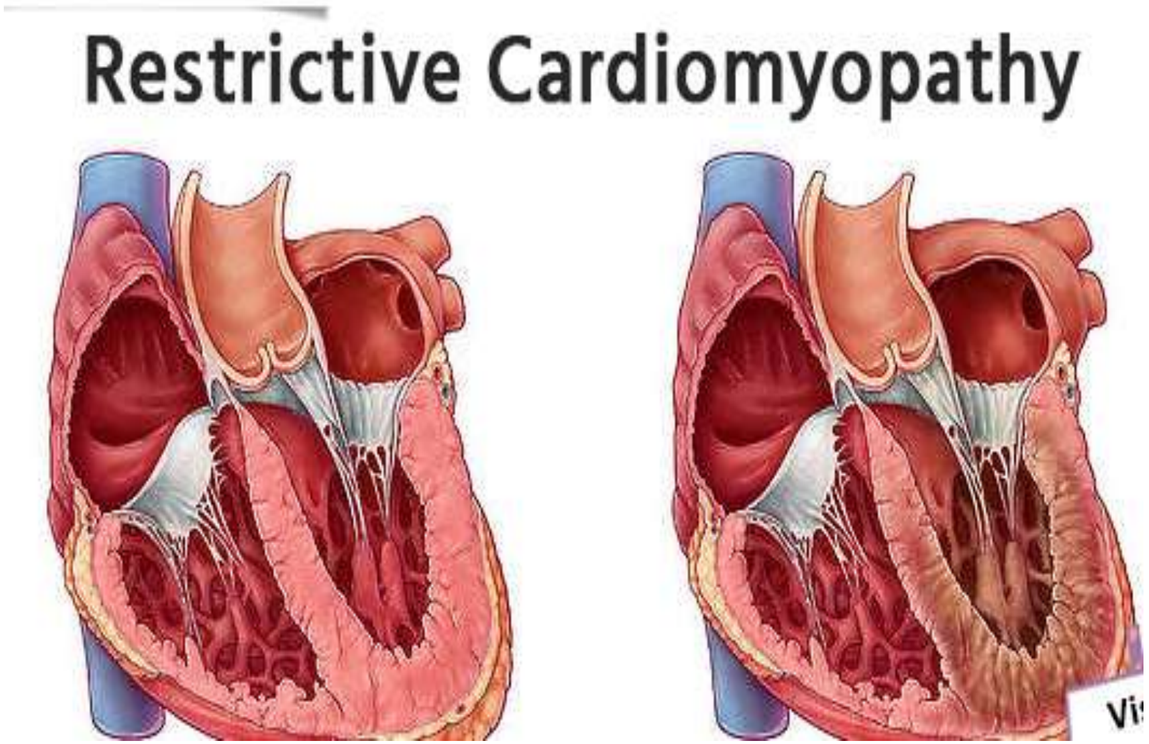
DEPARTMENT : DEPARTMENT OF CARDIAC TECHNOLOGY

SUBJECT : ECHOCARDIOGRAPHY

TOPIC : ECHO ASSESSMENT OF RESTRICTIVE CARDIOMYOPATHY

RESTRICTIVE CARDIOMYOPATHY

- Definition
- Epidemiology
- Classification
- Genetics & Etiology of RCM
- Clinical characteristics
- Diagnostic evaluation
- Management



DEFINITION

- Disease of heart muscle with restrictive physiology
- Stiffened or non-compliant ventricle → severe diastolic dysfunction & ↑ ventricular filling pressures
- Normal systolic function
- Normal or ↑ wall thickness (in infiltrative causes)
- Normal LV cavity & biatrial enlargement

CLASSIFICATION –RCMP

- Based on etiology – primary & secondary
- Primary include idiopathic, familial & endomyocardial fibrosis
- Secondary forms include infiltrative diseases, storage disorders, metastatic & iatrogenic causes

ETIOLOGIES :

GENETICS OF RCM

- Gene Mutations associated with RCM phenotype include **Sarcomeric & Non-sarcomeric protein defects**
- Modes of inheritance: AD, AR, X-linked & denovo (rare)
- Many families have common gene defect but different phenotypic expression (eg: *TNNI3 – Troponin I mutation – express RCM and HCM phenotype)*

IDIOPATHIC RCM

- ✓Seen from infancy to adulthood
- ✓Poor prognosis especially in children
- ✓Genetically determined or familial in most cases
- ✓Atrial fibrillation is common

Cardiac amyloidosis:

- Infiltrative cardiomyopathy
- MC types associated with cardiac amyloidosis – AL; ATTRw; ATTRm and Localised atrial amyloidosis
- Clinical pattern, prognosis & management depend on type *AL type* has clinically significant cardiac involvement in upto 75%
- Rapidly progessive heart failure along with systemic disease (survival time ≤ 9 m)
- *Familial amyloidosis (ATTRm)* –autosomal dominant;
- Involve both heart and peripheral nervous system
- Young pts – predominant peripheral nervous system;
- Middle age pts –predominant cardiomyopathy

- SARCOIDOSIS
- ENDOMYOCARDIAL FIBROSIS
- ENDOCARDITIS
- CARCINOID HEART DISEASES

PATHOPHYSIOLOGY

- ↑Ventricular stiffness & compliance→ impaired relaxation & restrictive filling → marked pressure increase with small vol. rise → diastolic dysfunction
- Rapid rise in ventricular filling pressures during exercise → limited stroke vol. → syncope/ angina / DOE
- Atrial enlargement → development of atrial arrhythmias & secondary AV valve regurgitation
- Thromboembolic complications seen with or without AF

CLINICAL FEATURES

- Symptoms of heart failure – DOE, exercise intolerance, fatigue, syncope and angina (rare)
- Physical findings: predominant right heart failure features –
- Raised JVP with prominent X and Y descent (Kussmauls sign)
- Irregular pulse- atrial fibrillationApex beat – usually palpable, may be mildly displaced
- Heart sounds- S1, S2 normal, S4 gallop frequently heard except in amyloidosis
- Murmurs- usually not heard, MR & TR seen in some
- Hepatomegaly, ascites and marked pedal edema

DIAGNOSIS EVALUATION

ECG :

✓Right and left atrial enlargement (MC); Non-specific ST-T changes; conduction abnormalities

✓ Atrial fibrillation is common;

✓Low voltage limb leads with Pseudo-infarction pattern in inferior & septal leads seen in cardiac amyloidosis

DIAGNOSIS EVALUATION

HOLTER:

- Useful esp. in children for Rhythm evaluation & ST segment analysis
- Studies done by Rivenes et al and Greenway et al reported rate related ST segment depression in patients with anginal episodes/ arrhythmias
- On contrary, Hayashi et al didn't find any coronary abnormalities by catheterization and perfusion defects by exercise testing
- Approximately 15% of pediatric population had arrhythmias & conduction disturbances
- Atrial flutter was MC reported followed by high degree second and third degree heart block
- Longer PR interval and longer QRS duration were associated with acute cardiac events

DIAGNOSIS EVALUATION

ECHOCARDIOGRAPHY:

Intial step in the diagnosis

2D ECHO:

Typical features in RCM incudes :

✓Normal RV and LV EF;
✓Normal RV and LV chamber volumes & wall thickness ;
✓(exception → wall thickness increased in infiltrative conditions)
✓Biatrial enlargement

DIAGNOSIS EVALUATION

ECHOCARDIOGRAPHY:

Doppler ECHO findings (s/o restrictive filling parameters) are as follows:

✓Increased E/A ratio > 1.5
✓Decreased mitral deceleration time (DT < 120ms)
✓Decreased IVRT (isovolumetric relaxation time)
✓Decreased PVs/PVd ratio (pulmonary venous flow velocities)
✓Augmented atrial reversal velocity (PVAr)

❖ *Doppler tissue imaging (DTI):*

✓Reduced mitral anular velocities (e')
✓Increased E/e' ratio

❖ *Hepatic vein doppler:*

✓Increased diastolic forward flow reversal with inspiration

DISEASES SPECIFIC ECHO FINDINGDS

- *Amyloidosis :* ↑ RV and LV wall thickness
- very *fine granular or scintillating echobright* appearance of myocardium with preserved LVEF
- Diffuse thickened AV valves with severe biatrial enlargement (owl eye appearance)
- *Doppler tissue imaging*- severly impaired longitudinal LV systolic function with normal EF
- *Speckle tracking* – regional longitudinal dysfunction with apical sparing
- *Haemochromatosis:*

✓LA, LV dilatataion
✓Normal LV wall thickness
✓Global hypokinesia
✓Initially restrictive pattern progressing to systolic dysfunction

DISEASES SPECIFIC ECHO FINDINGDS

Fabrys disease:

- Concentric LVH
- Mitral leaflet thickening with significant MR
- *Endomyocardial fibrosis:*
- Hallmark feature- *formation of diffuse thrombi along the endocardium in apices*

<u>Thrombus obliterating the ventricular cavity</u>

- Retraction of AV valves → incompetence

Endomyocardial fibrosis:

- Dilated hypocontractile LV and RV
- Absence of myocardial edema
- Normal myocardial perfusion
- Diffuse subendocardial LGE of LV/RV with/without thrombus
- Obliterative LV or RV apex

DISEASES SPECIFIC ECHO FINDINGDS

❖ Cardiac sarcoidosis:

✓Multifocal intense LGE often in septum inferolateral valve of LV, right atrium and RV free wall

✓RWMA with hypocontractile LV/RV

✓Patchy bright regions s/o *myocardial edema*

✓Normal myocardial perfusion

❖ *Haemochromatosis:*

✓Hypocontractile LV with dark myocardium

✓Absence of myocardial edema

✓Normal myocardial perfusion

✓Normal LGE mapping

ENDOMYOCARDIAL BIOPSY

- Important role in the diagnostic evaluation of diseases with restrictive physiology
- Class II a recommendation by AHA/ ACC
- RV biopsy is most commonly done
- Useful in identifying the exact etiology of RCM for targeted therapy

MANAGEMENT
GENERAL TREATMENT PRINCIPLES:

- Relieve congestive symptoms and avoid hypotension
- Diuretics
- Salt restriction (2-4 gms/day), fluid restriction to less than 2 litres
- Rx of AF with *rhythm control* rather than rate control improves diastolic filling
- No pharmacological treatment prolongs survival

DISEASES SPECIFIC TREATMENT:

❖ *Cardiac amyloidosis*:

✓AL amyloidosis- chemotherapy, autologous stem cell transplantation and monoclonal antibodies (daratumumab) targeted against plasma cell dyscrasias

✓Beta blockers, ACEI/ ARBs poorly tolerated

✓CCBs contraindicated (worsens heart failure)

✓High degree AV block may require permanent pacemaker- biventricular pacing preferred*Familial amyloidosis (ATTRm):*

✓Liver/ heart-liver transplantation and tafamidis (TTR stabilizer)

✓*Senile systemic amyloidosis (ATTRw):*

✓Gerneral treatment principles + tafamidis

- ***Cardiac sarcoidosis***:

✓In patients with heart failure Prednisolone 1mg/kg tapered over several months is recommended

✓High degree AV block require ICD- pacemaker implantation

✓Heart transplantation in treatment refractory pts

- ***Fabry's disease*** : enzyme replacement will reverse phenotype
- ***Hemochromatosis*** : iron chelation therapy ; frequent phlebotomies
- ***Carcinoid heart disease***: hepatic mets debulking ; use of octreotide – tumor shrinkage; advanced valve disease is surgically corrected
- ***Hypereosinophilic syndromes***: early stage-steroids are useful;

advanced valve disease – surgically corrected

THANK YOU