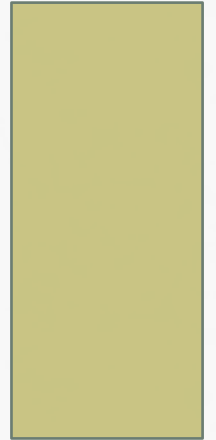


RESTRICTIVE CARDIOMYOPATHY

DR. DEEPTHI.G



HISTORY

- 10 yr old female by name y. Neeraja came with chief c/o
- Shortness of breath since 6 years associated with occasional cough & fever

HISTORY

- SOB initially noted at the age of 2 years as chest in drawing while feeding gradually progressive as she has difficulty in playing with the same age group people.

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HISTORY

- Cough since 6yrs
episodic
not associated with sputum
last for few days
relieved by taking medications from local
shop

HISTORY

- h/o fever present episodically during episodes of cough,
 - associated with chills
 - lasts for 2-3 days
 - relieved with medication.
- h/o difficulty in weight gain is present

HISTORY

- No h/o
 - cyanotic spells
 - seizures
 - syncope
 - vomitings
 - diarrhoea
 - altered sensorium

HISTORY

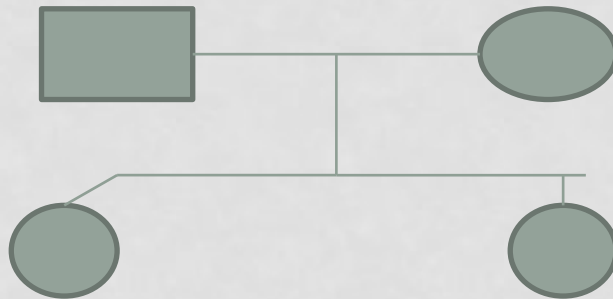
- With these complaints patient went to a local hospital where she was diagnosed as having a heart disease at the age of 10 yrs.
- No h/o bronchial asthma, tb, epilepsy in the past

HISTORY

- Ante natal h/o –
took antenatal medications and vaccinated regularly
no episodes of fever with rash
- Natal h/o -
NVD, home delivery,
conducted by anganwadi worker
baby cried immediately after birth
- Post natal h/o –
vaccinated up to date

HISTORY

- Family history –
- non consanguineous marriage.
- elder child
- no similar complaints in the other child



PHYSICAL EXAMINATION

- Height – 112 cms
- weight – 18 kgs
- Temp – afebrile
- pr – 98/min
- BP – 100/60 mm
- JVP – elevated with prominent X and Y DESCENT.
- p/A – hepato spleno megaly present

INVESTIGATION

- Hb%:14.0
- TLC:11,000 cells/cumm
- Platelet:2.44 lakhs/cumm
- ESR:15
- Normocytic normochromic with eosinophilia

- Mantoux: negative
- USG ABDOMEN – hepatomegaly s/o congestion
- Viral markers – negative
- BCA - normal
- CHEST X RAY – CARDIOMEGALY{prominent rt atrial enlargement}.

ECHO

- Dilated RA
- Mild MR ,Mild TR ,No PAH
- Normal ventricular sizes
- Fair LV systolic function
- Mild PE
- Dilated IVC with no respiratory variation
- Features s/o restrictive cardiomyopathy ,dilated RA ,NO PAH, NO PERICARDIAL THICKENING

CATH DATA

- Right heart study done
- aorta 100/50
- PA 40/15/19
- RA 16/6
- RV 45/16
- RVEDP 16
- LV 90/18
- LVEDP 18

CARDIAC MRI WITH CONTRAST

- Changes in RV, LV myocardium s/o restrictive cardiomyopathy
- Moderate pericardial effusion with no pericardial thickening / enhancement
- Moderate TR, mild PAH
- Mild ascites ,no pleural collection
- Enlarged mediastinal lymph nodes, largest measuring 12mm

- Whether it is RESTRICTIVE CARDIOMYOPATHY
or
CONSTRICTIVE PERICARDITIS

DISCUSSION

- Restrictive cardiomyopathy accounts for 2-3 % of all cardio myopathies in children, present with symptoms of heart failure or syncope
- Myocardial causes of RCMP:
- Infiltrative:
 - 1.amyloidosis
 - 2.sarcoidosis
 - 3.hemochromatosis
 - 4.lysosomal storage disorder

- Non infiltrative:
 1. idiopathic
 2. familial
 3. post transplant
 4. diabetic cardiomyopathy

Endomyocardial causes:

1. EMF
2. hyperesinophilic syndrome

THANK U