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CASE REPORT OF RARE ASSOCIATION OF JUVENILE RHEUMATIC MITRAL STENOSIS WITH PERIMEMBRANOUS VSD

Pediatrics

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ABSTRACT

Rheumatic mitral stenosis (MS) is one of the most common condition seen in the developing world. But the presence of mitral stenosis with severe symptoms at a very young age is rare. We report a case of an unusual association of a moderate perimembranous ventricular septal defect(VSD) with juvenile rheumatic mitral stenosis leading to severe pulmonary hypertension and heart failure in a 7 year old child. The association of congenital ASD with acquired Rheumatic MS (LUTEMBACHER SYNDROME) is a well known entity. But, the association of a congenital VSD with rheumatic MS is very rare.

KEYWORDS

Rheumatic mitral stenosis, perimembranous VSD, Lutembacher syndrome.

INTRODUCTION:

Rheumatic mitral stenosis (MS) is one of the most common condition seen in the developing world. But the presence of mitral stenosis with severe symptoms at a very young age is rare.

CASE REPORT: A 7 year old male child born out of 2nd degree consanguinous parents belonging to upper lower class presented with symptoms of Dyspnoea, Paraoxysmal nocturnal dyspnea and Orthopnoea. There was history of hospitalisation for LRTI at 4th month of age and was diagnosed as having acyanotic congenital heart disease(VSD) and was advised repair. The child had recurrent episodes of lower respiratory tract infections and failure to thrive. Antenatal , natal and family history -not significant.

ANTHROPOMETRY-Weight-14 Kg (< 3 rd centile),
Height-108 cm (< 3 rd centile),
Head Circumference- 45 Cm (< 3 rd centile).

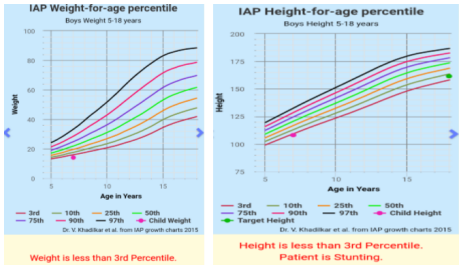


figure 1: The weight and length of the child was below 3rd centile according to IAP growth charts.

VITALS are within normal limits.

SYSTEMIC EXAMINATION:

CVS- Precordial bulge with precordial and epigastric pulsations present. On Auscultation, Mitral area-Loud first heart sound with opening snap and grade 4 Mid Diastolic Murmur heard. Pulmonary area-Loud pulmonic component of second heart sound and Ejection systolic murmur heard. Tricuspid area – pan systolic murmur is heard with more intensity in left 3rd and 4th parasternal area.
RS – bilateral basal crepitations heard.

INVESTIGATIONS:

CBC suggestive of leucocytosis, ASO titres were raised (400 Todd units) ,
LFT , RFT and SERUM ELECTROLYTES were normal .
ECG suggestive of RAD, RVH and Right ventricular strain pattern.

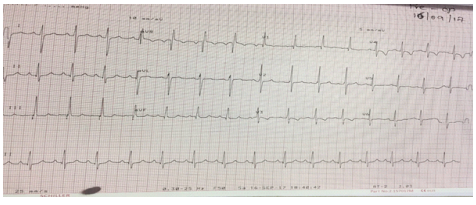


Figure 2: ECG suggestive of RAD, RVH and Right ventricular strain pattern.

CHEST X-RAY PA VIEW suggestive of Cardiomegaly with right ventricular enlargement with pulmonary plethora.

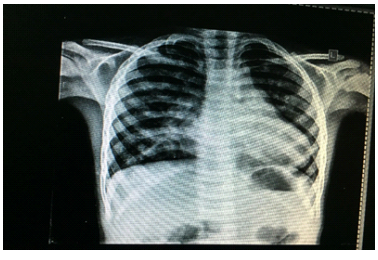
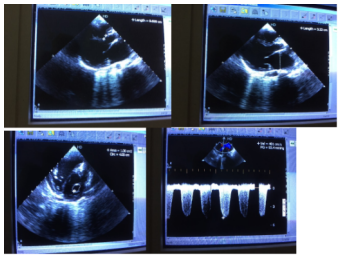


Figure 3: CHEST X-RAY PA VIEW suggestive of Cardiomegaly with right ventricular enlargement with pulmonary plethora.

2D Echo- Moderate peri membranous VSD (0.7 cm2),
Moderate mitral stenosis (MVA -1.1 cm2) with no mitral regurgitation, Thickened mitral valves seen, Dilated LA (3.2cm) & RA, Severe TR with Severe PAH ,RVSP- 102mm Hg,TJV-4.8m/sec, EF-56%.



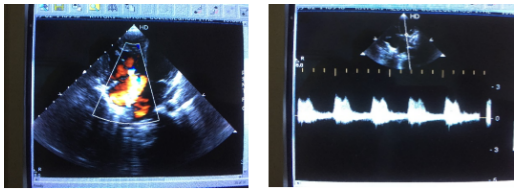


Figure 4: 2D Echo images of the child showing perimembranous VSD with severe mitral stenosis.

TREATMENT: The child was initially treated with Diuretics, Digoxin and antibiotics. Later child was operated under cardiopulmonary bypass at our hospital. VSD was closed with Dacron patch and open Mitral valvotomy was performed

INTRAOPERATIVE FINDINGS: Large sub aortic VSD, mitral commissural fusion and very severe PAH.

FOLLOW-UP: Child had good post operative recovery and was discharged on diuretics and Inj.Benzathine penicillin prophylaxis(3rd weekly).He was asymptomatic after 1 month of follow up.

DISCUSSION: The incidence of congenital heart disease is about 8 per 1000 live births. The prevalence of Rheumatic heart disease in school-aged children is in the range of 2-11 per 1000(1). (2). Group A β-haemolytic streptococcus is the major etiological agent for causing acute rheumatic fever through autoimmune process (3). The damage to cardiac valves caused by the immune mediated process results in RHD. The commonest valve affected is mitral valve (4).Developmental defects at early embryonic period results in CHD. Approximately 10% of the patients with juvenile rheumatic MS present before 12 years of age and associated with rapid haemodynamic progression with severe symptoms (5). As both disease process are different and had varied incidence, the association of both these conditions can be considered as uncommon. The exact Etiology for occurrence of this association is not known. This association may be a mere coincidence or CHD might predispose to RHD Our case report discussed here is probably one of the youngest case of juvenile Rheumatic MS with VSD supported by intraoperative findings.

CONCLUSION: The association of congenital ASD with acquired Rheumatic MS (LUTEMBACHER SYNDROME) is a well known entity. But, the association of a congenital VSD with rheumatic MS is very rare. This association emphasize the need for careful evaluation of children with CHD for later development of rheumatic heart disease or coexistent RHD. This association causes rapid hemodynamic deterioration and alters the management plan.

Conflict of Interest:
There is no conflict of interest among the authors.

Funding Statement:
There is no funding for the case report.

Ethical Approval:
Necessary approval was taken from the Institution and the patients for carrying out this work

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