Abstract: Rheumatic fever (RF) and rheumatic heart disease (RHD) continue to ravage millions of people around the world. Children and adolescents in developing countries are especially susceptible, due to overcrowding, poor sanitary conditions and low access to healthcare facilities. Such circumstances sometimes result in severe and early onset complications that are uncommonly seen in developed countries. We present a case of severe RHEUMATIC MITRAL STENOSIS in an Indian boy as young as 13 years of age.

Keyword: mitral stenosis, rheumatic heart disease

CASE HISTORY: 13 year old Indian male, first born of a non-consanguineous couple, presented with a 2 month history of restricted general activity and progressive exertional dyspnoea. General examination revealed a thin young boy 140 cm in height weighing 25 kg, with no structural abnormalities. Cardiovascular examination revealed a regular heart rate of 82/min and BP 100/60 mmHg with an elevated JVP. The apex beat was tapping in character and located in the 5th intercostal space (medial to the mid-clavicular line). A diastolic thrill was present at the apex. Additionally, a Grade 2 left parasternal heave (with P2 palpable in the 2nd left intercostal space) accompanied by epigastric pulsation was noted.

On auscultation, there was a low pitched rough rumbling mid-diastolic murmur (Grade 4/6) in the mitral area with a loud first heart sound and opening snap. Tricuspid regurgitation and a Grade 3/6 ejection systolic murmur (pulmonary region) with a loud P2 were also noted. All other systems were normal. Investigations showed Hb at 13 gm/dL (male 13-18 gm/dL) and a random blood sugar of 98 mg/dl (70-110 mg/dl). Serum electrolytes, renal and liver function tests were normal. Abdominal ultrasound was also normal.

ECG shows right axis deviation, right atrial enlargement and right ventricular hypertrophy.

Chest X-ray shows left and right atrial enlargement, left main pulmonary artery prominence.

Echocardiography (Fig 3a, 3b, 3c): Anterior and posterior mitral leaflets are thickened. Posterior mitral leaflet has restricted mobility. Grade 2 sub-valvular disease is present. Mitral valve shows severe mitral stenosis mitral valve area reduced to 0.65 cm², mitral mean gradient is 14 mmHg, peak gradient is 30 mmHg. Severe pulmonary hypertension with pulmonary gradient of 80 mmHg. Tricuspid valve shows moderate tricuspid regurgitation. Aortic valve thickened. No evidence of infective endocarditis, left atrial thrombus or atrial fibrillation.

Echo shows Mitral Valve Area reduction to 0.65 cm².

Echo illustrates: 1) Thickened Anterior and Posterior Mitral leaflet. 2) Hockey stick appearance of Anterior Mitral leaflet.
Echo shows increased diameter of the Left Atrium
The primary diagnosis was severe mitral stenosis with severe pulmonary hypertension (of rheumatic etiology) with accompanying heart failure. The boy was treated with salt restriction, IV frusemide and oral digoxin; Oral penicillin was also commenced for secondary rheumatic fever prophylaxis. Patient improved symptomatically over 5 days and was discharged with regular follow-up.

DISCUSSION Rheumatic fever results from an autoimmune response to infection with group A beta haemolytic streptococcus. Although the acute illness causes considerable morbidity, and some mortality, the major clinical and public health effects derive from long-term damage to the heart valves, i.e., rheumatic heart disease (RHD). Over the last century, with improved living conditions including better nutrition, hygiene and increased access to medical care, acute rheumatic fever (ARF) and RHD have become rare entities. But in developing countries such as India, it remains the commonest form of valvular heart disease. Between 1940 and 1983, the prevalence rate for RHD varied from 1.8 to 11 per 1000 (national average 6 per 1000), while between 1984 and1995 the rate varied from 1 to 5.4 per 1000. (1)During the same period, the prevalence of rheumatic fever ranged from 0.06 to 5.01 and 0.32 to 0.54 per 1000, respectively. Due to the diverent methods of collecting the data, it is not possible to be certain that these gures represent a fall in the prevalence of RHD. (1)By comparison in developed nations, the prevalence of RHD in children aged between 5–15 years is below 0.5 per 1000. (1)Carditis can occur in the acute form of rheumatic fever and can progress to rheumatic heart disease. The inflammatory process in the valve leaflets is thought to be initiated by cross-reactivity between streptococcal antigen and the valve tissue. Although the incidence of rheumatic heart disease is variable after an episode of acute rheumatic fever, approximately 50 percent of those with evidence of carditis develop organic valvular disease.

(2) Rheumatic mitral stenosis (MS) has a delayed onset compared to rheumatic mitral regurgitation (MR). In a report of patients with surgically treated rheumatic mitral valve disease, most cases with pure MR occurred in those under 30 years of age, while pure mitral stenosis and mixed mitral stenosis and regurgitation increased progressively over time with a peak prevalence over age 30 (3) In most patients, mitral stenosis is a progressive disease. Progression is slow in asymptomatic patients, but becomes more rapid after the onset of symptoms. The rate of progression of MS varies across geographical areas (4). The mean rate of progressive valve narrowing is approximately 0.1 cm2/year in North American series, but there is appreciable inter patient variability (5). The mean interval between rheumatic fever and the onset of symptoms was 16.3 years in a prospective study from Germany. (2)

In developing countries, mitral stenosis progresses rapidly, and may lead to symptoms in younger patients such as young adults or teenagers. But even children less than 5 years of age have been reported to be symptomatic in some countries. (6) Such quick rates of progression have been attributed to the ineffective use of antibiotics and/or increased virulence of the Streptococcus organism, which can cause a more severe primary rheumatic insult. (3) The ensuing acute inflammatory process can lead to comissural adhesion prior to the more common degenerative sequelae. (3) Other causes include repeated and ongoing infection possibly driving the valvular inflammatory response.

. It is therefore not uncommon in some countries to see symptomatic MS in patients less than 20 years of age (3), compared to the typical development of symptoms between the ages of 30 and 50 in developed nations (7). One of the possible reasons for the early onset RHD MS in our patient may be severe primary rheumatic insult due to ineffective use of antibiotics or increased virulence of Streptococcus. (3)Consequentially commissural adhesions may have developed prior to the common degenerative disease, hastening the development of symptomatic disease. Other possibilities include recurrent rheumatic fever because of high endemicity of the disease. Interestingly the patient and his family denied a history of rheumatic fever; possibly due to unrecognized disease at an age less than 5 which is unusual (usually affects 5-15yr olds). (8) In summary, cases of such advanced mitral valve disease in a child as young at 13 years of age have rarely been reported previously. It is evident that further research needs to be done to ascertain the prevalence and aetiology of early onset symptomatic RHD. Public education regarding the disease and its effects may help raise detection rates and compliance with regular follow-up. Research and education combined with improving hygiene and better health care, may contribute to reduced incidence of this disease among children and young adults in India.

REFERENCES
1) Padmanavati S. Present status of rheumatic fever and rheumatic heart disease in India. Indian Heart J 1995;47:395–8
7) Selzer A, Cohn KE. Natural history of mitral stenosis: a review. Circulation 1972; 45:878