

# Juvenile Mitral Stenosis

# Introduction

- Earlier documentations of Rheumatism came from the West
- Rheumatic Heart Disease was regarded a disease of the temperate climates
- In fact Rogers<sup>1</sup> reported that except for a single case, he did not find RHD in 4800 postmortem records, over 37 years in Calcutta
- Eventually in the early 20<sup>th</sup> century, Wig et al<sup>2</sup> first and then others described a significant burden of the disease in India (and surrounding countries as well)
- Kutumbiah<sup>3</sup> in 1940 reported on the disease in adolescence and childhood

# Introduction

- Over the years, several hospital based studies documented the burden on Rheumatic Heart Disease in patients around the country
- Rheumatic Fever in India had been identified as a severe disease with multivalve involvement and congestive heart failure

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## AN APPRECIATION OF MITRAL STENOSIS\*

### PART I. CLINICAL FEATURES

BY

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# Paul Wood series

- 150 cases planned for surgery and another 150 unsuitable for surgery were studied

# Paul Wood series

- While other authors felt that incidences of individual valve involvement may have been determined by frequency with which each valve was vascularized
- Wood felt this had more to do with the degree of stress to which each valve was subjected



# Paul Wood series

- Previous history of the rheumatic fever in 68% of patients
- Past history of Rheumatic Fever was obtained with highest frequency in patients with Multivalve disease > MR > Pure MS
- Based on this he felt that pure mitral stenosis represented the least florid form of rheumatic heart disease
- Among patients with past history of the rheumatic fever

45.7% - single attack

31.5% - recurrent attacks

12.5% chorea alone

4.8% both chorea and rheumatism

and 5.5% had had only subacute rheumatism or growing pains



# Paul Wood series

- Cases with significant mitral incompetence or multiple valve lesions were ten times more likely to have had recurrent rheumatic fever than isolated chorea;
- On the other hand; in relatively mild cases of pure mitral stenosis a previous history of chorea alone was twice as common as recurrent rheumatic fever
- This strengthened the suggestion that pure MS was the mildest form

# Paul Wood series

TABLE III.—*Life History*

	Latent period			Duration of Symptoms				From Onset Symptoms to Total In-capacity (Years)
	Age Initial Attack	Latent Period (Years)	Age Onset Symptoms	Grade 1 (Years)	Grade 2 (Years)	Grade 3 (Years)	Grade 4 (Years)	
Mitral stenosis	12	19	31	2.7 (0-17)	2.7 (0-18)	1.94 (0.5-11)	2 (0.5-4)	7.3
Mitral incom- petence	12.5	20	32.5	(0-9)	(0-5) 1.1	2.25 (0-7)	(0.3-3) 1	5.4

# Paul Wood series

- Functional Class

Class II – 19%

Class III – 41%

Class IV – 39%

- All symptomatic patients had elevated left atrial pressures (except 1 who was operated for recurrent embolism)
- Orthopnoea was present in 53% patients in the whole series (69% in surgical group, 30% of medical group, 44% patients with MR)

# Paul Wood series

- To assess which factors governed presence of symptoms due to pulmonary venous congestion, they studied patients with orthopnoea/PND with those without
- Degree of stenosis was not related to functional behavior
- Rather Pulmonary Vascular Resistance in patients with PND was half that of those without PND
- Lower PVR did not protect the lungs adequately

# Paul Wood series

- Even in case of patients with frank Pulmonary Edema

Age was lower

More often Female

Lower PVR

More often sinus rhythm

Somewhat lesser stenosis than those without pulmonary edema

LA pressure was high in both groups (with or without pulm edema)

# Paul Wood series

- AF occurred more often in older patients than in younger patients
- Mean age
  - AF – 41
  - Sinus Rhythm - 33.6 years

# Juvenile Rheumatic Stenosis and Dr. Sujoy B Roy

- Described the unusual features of Juvenile MS (and coined the term) in an article and a presentation at the 7<sup>th</sup> International Congress on Diseases of Chest (New Delhi, February 1963)
- Published in *The Lancet* (*The Lancet 1963; 2:1193-6*)

## JUVENILE MITRAL STENOSIS IN INDIA\*

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# Roy series

- Of a total of 2383 patients seen in the cardiology clinic over 4 years, 754 patients had Rheumatic Heart Disease
- 171 patients were below the age of 20  
Patients with significant MR or aortic valve involvement were not included in the study
- 108 patients had pure Mitral stenosis
- 66 M, 42 F



# Roy series

- Right heart cath was done in 62 of these patients
- 23 of these patients had operations on the mitral valve
- At surgery, tissue from the LA appendage, parietal pericardium and left lower lobe of lung was taken

# Roy series

- 71 (66%) of patients had a history of at least 1 episode of ARF
- 30 (28%) had more than one attack (history wise)
- Diagnosis of rheumatic fever was based on a history of migratory polyarthrititis (more often polyarthralgia), fever and manifestations of carditis
- Chorea and Subcutaneous nodules were infrequent (<3%)
- Erythema Marginatum was conspicuously absent

# Roy series

- Interval between the first attack of rheumatic fever and the onset of symptoms varied - a few weeks to several years
- **50 (70%) of the 71 patients** with a history of rheumatic fever had symptoms within 5 years of the first attack

TABLE I—CLINICAL FEATURES IN 108 PATIENTS WITH JUVENILE MITRAL STENOSIS

Feature	No. of males (66)	No. of females (42)	Total
<i>History of rheumatic fever</i>	45	26	71
<i>Dyspnoea:</i>			
Mild	7	4	11
Moderate	26	20	46
Severe	26	13	39
<i>Paroxysmal nocturnal dyspnoea</i>	10	7	17
<i>Haemoptysis</i>	15	14	29
<i>Angina</i>	7	6	13
<i>Embolism</i>	1	1	2
<i>Congestive heart-failure</i>	32	17	49
<i>Rheumatic activity</i>	12	12	24
<i>Asymptomatic</i>	7	3	10

TABLE II—VALVULAR DISEASE IN 171 PATIENTS WITH JUVENILE RHEUMATIC HEART-DISEASE

Valvular lesion	No. of patients affected
Mitral stenosis	69 (40%)
Mitral stenosis with slight regurgitation	39 (33%)
Mitral stenosis with moderate regurgitation	14 (8%)
Mitral regurgitation	12 (7%)
Mitral and aortic-valve disease	31 (18%)
Aortic regurgitation	6 (3%)

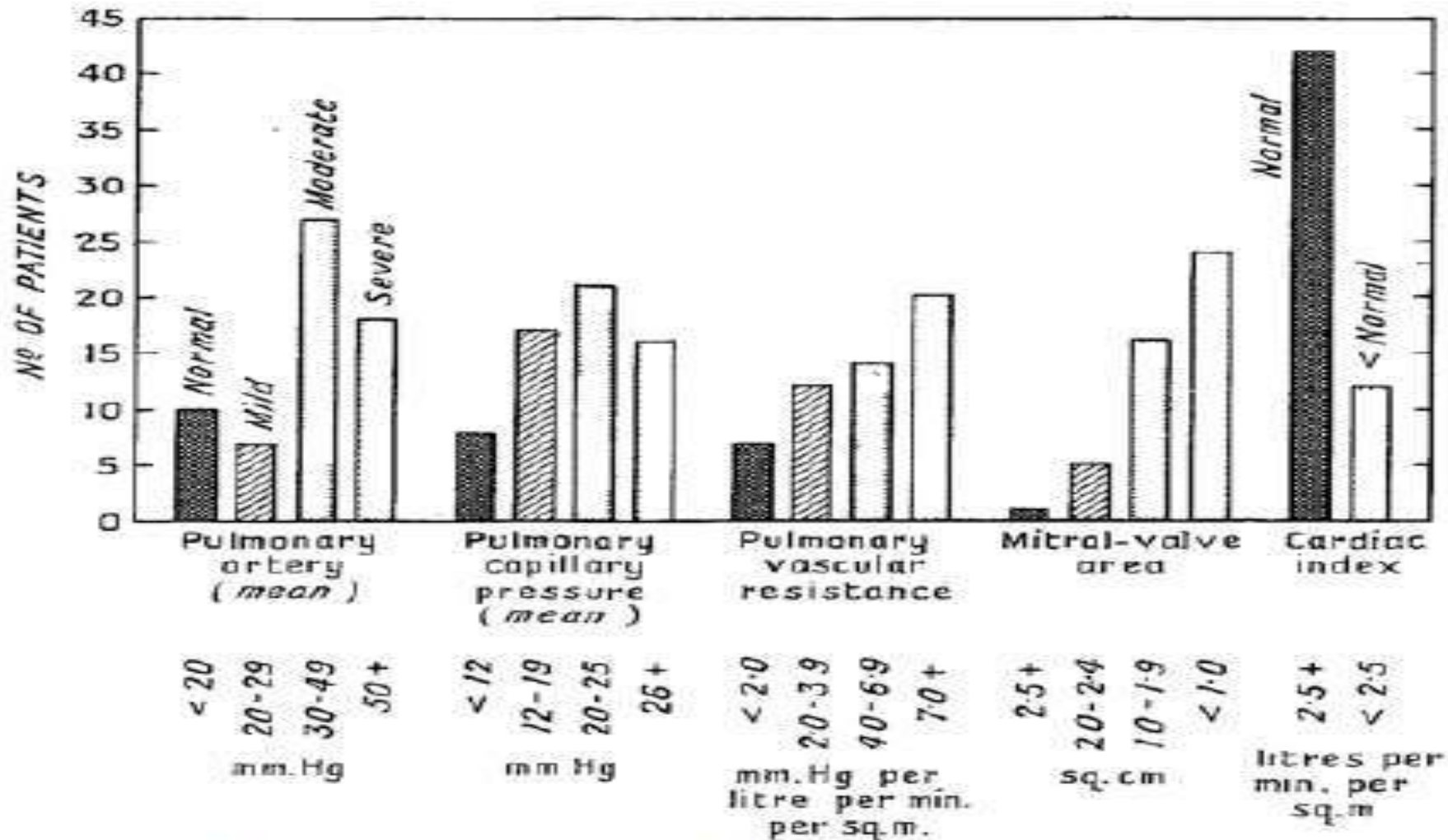
# Roy series - ECG and X rays

- Routine 12-lead electrocardiograms showed moderate right ventricular hypertrophy (R/S ratio in lead V1 greater than 1) in 38 patients and severe (R/S ratio in lead V1 greater than 5) in 26 patients.
- The prime R in lead V1 exceeded 12 mm in 13 patients
- Radiological changes consistent with moderate to severe degrees of pulmonary hypertension were observed in 68 patients

# Roy series - Rheumatic Activity

- Rheumatic disease was active in 24 patients (based on Jones' criteria)
- But if congestive cardiac failure was considered evidence of carditis and therefore active disease, total number was 49

# Roy series - Hemodynamic data



**Fig. 1—Haemodynamic findings in 62 cases of juvenile mitral stenosis.**



# Roy series - Hemodynamic data

- In over two-thirds of the catheterised patients the resting mean pulmonary arterial pressures were considerably raised pressures
- 60% of the patients the resting pulmonary arterial wedged pressures exceeded 20 mm hg
- Cardiac output, on the other hand, was normal in 80% of the patients
- The pulmonary vascular resistance was grossly abnormal in almost two-thirds of the patients
- The estimated mitral-valve area was less than 1 sq. cm in most

# Roy series - Surgical Findings

- Calcification and mural thrombi were detected in only 1 patient
- In 17 of the 23 patients, the pulmonary arterial trunk was large and the aorta was relatively small

# Roy series - Histopathological changes

- Small muscular branches of the pulmonary artery accompanying the terminal and respiratory bronchioles and in the pulmonary arterioles and venules there was pronounced medial muscular hypertrophy with either concentric or eccentric thickening of the intima
- Severe narrowing of the lumen was the rule
- Rigidity and sclerosis of the alveolar capillaries with thickening and duplication of their reticular framework were prominent features

# Summary

- Isolated mitral stenosis in patients below the age of 20 with rheumatic heart-disease is common in India
- M>F (c.f adults)
- Erythema marginatum is not seen, S/C Nodules rare
- Congestive heart-failure is a frequent finding in this age-group
- Atrial fibrillation uncommon in this subset

# Summary

- High prevalence of severe pulmonary hypertension with gross pulmonary vascular changes seen in this group thought to suggest
  - a hypersensitive reaction of the pulmonary vasculature to a fulminating rheumatic process,
  - or a tissue response to multiple overt attacks of rheumatic fever

Reale et al 1963

**Mitral stenosis in childhood:  
Clinical and therapeutic aspects**

*Attilio Reale, M.D.\**

*Ciriaco Colella, M.D.*

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*Rome, Italy*

# Reale series

- Over 13 years, 54 patients between ages 8-15 admitted with Mitral stenosis were studied
- 17M, 37F
- 48 were subjected to commissurotomy
- Mean age 12.7 years
- They compared the subset of young patients to an older subset treated at the same institute during the same period

# Reale series

- In 21 patients, (39 percent) had a history of typical acute rheumatic fever, with repeated episodes reported in 7 (13%)
- Average age of first bout of ARF approx. 8, average age at onset of cardiovascular symptoms 10, with average interval between onset of ARF and development of symptoms of 2 years (minimum 6 months, max 10 years)
- Younger patients more often presented with pulmonary edema, while older patients presented with hemoptysis and pedal edema



# Reale series

- Compared to the adults they noted similar female predilection for the disease
- However symptomatology was more often exertional dyspnoea in children, most children being in NYHA Class IV
- Hemoptysis and Pulmonary Edema were frequent in children but AF and peripheral emboli were not
- Higher frequency of RVH findings on ECG
- LA enlargement seen far less often on x ray in children than adults

# Reale series

- Hemodynamically, children had a tendency for higher pulmonary arterial pressures and total pulmonary resistance
- Though the authors felt that higher pulmonary capillary wedge pressures meant a relatively lower arterial-venous gradient and relatively unaltered pulmonary arteriolar resistance
- They felt this reflected elevated pressures due to downstream (valvular) obstruction and precapillary vasoconstriction, and that pulmonary arteriolar anatomic changes had not yet achieved significance
- As a result, PAH was rapidly reversible after the obstruction was relieved (documented by post op cath at 1 month) when compared to adults (slower reversal)

# Reale series

- Histologically too, they compared lung biopsies from a younger patient to that of an adult with lower (half) the PA pressure
- They showed that in the younger patient, though arteriolar muscular hypertrophy was noted, the intima was intact and the lumen was patent
- Where as in the adult patient with a lower (relatively) PA pressure, the lumen was collapsed (complete occlusion)

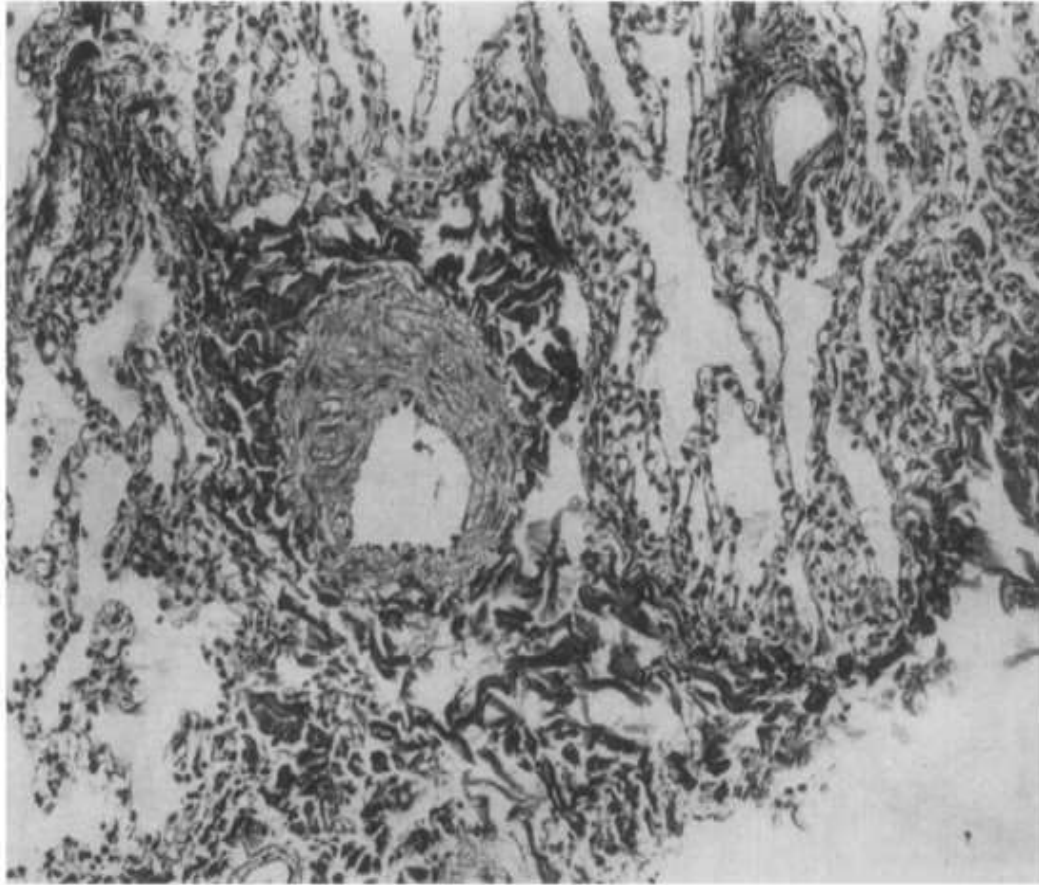


Fig. 5. Lung biopsy of Patient S.D., showing hypertrophy of a pulmonary arteriole with patent lumen and intact intima (Van Gieson stain).

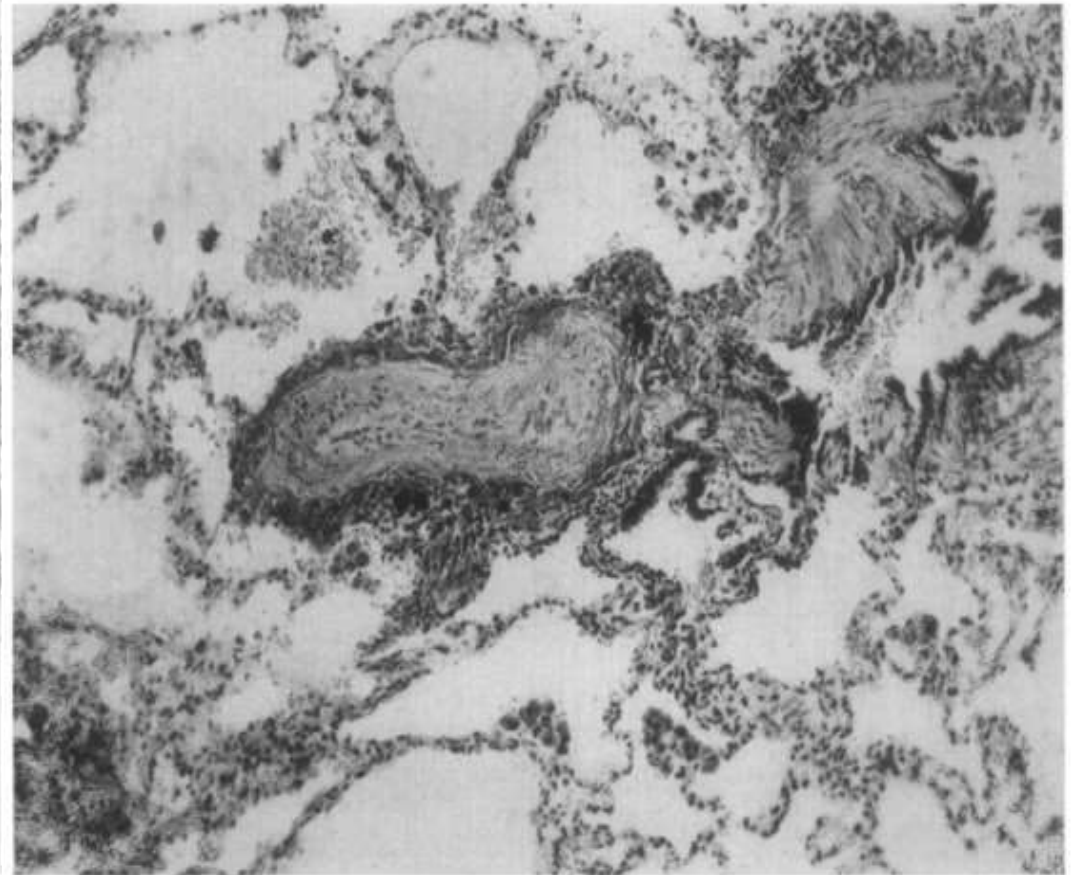


Fig. 6. Lung biopsy from an adult patient with mitral stenosis, showing complete obliteration of a pulmonary arteriole (Van Gieson stain).

# Reale series

- They suggested that the anatomic changes in the adult result not only from downstream obstruction but from the rheumatic process involving myocardium itself, from longstanding atrial dilatation resulting in fibrosis

# Summary and Conclusion

# The Problem

- Juvenile Rheumatic Heart Disease continues to be a significant problem (prevalence 1 - 5.4/1000 in children between 5-15)
- Early progression to severe valvular disease and severe symptoms occurs in a subset, resulting in manifestations at a younger age group

# A Different Entity

- Various case series of Mitral Stenosis in children have showed a different phenotype
- Specifically,

Shorter latency period

Early severe manifestations of disease

Predilection to Pulmonary edema, PVH

More rapid improvement in PA pressures after intervention

Far lower incidence of AF and systemic embolism



# Solution?

- With Rheumatic Heart Disease, prevention is the only real cure
- Primary and Secondary Prevention of utmost importance
- Early intervention to relieve valvular obstruction and correct incompetence is important