Epidemiological survey of rheumatic heart disease among school children in the Shimla Hills of northern India: prevalence and risk factors

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Abstract

Study objective – To determine the prevalence of rheumatic heart disease (RHD) and study the relationship of this disease to factors such as age, sex, housing, and socioeconomic status in Shimla town and the adjoining rural area.

Design – A cross sectional survey, carried out by a specially trained examiner in cardiology.

Setting – The study involved high risk school children (5–16 years of age) from Shimla town and the adjoining rural area of Kasumpti-Suni Block in the period 1992–93.

Subjects – A total of 15 080 children on the school register (8120 boys and 6960 girls) were examined generally and specifically for evidence of RHD.

Main results – Of the 15 080 children screened, the prevalence of rheumatic fever (RF)/RHD was 2.98 per thousand with no significant difference between the age groups of 5–10 and 11–16 years or in either sex ($p>0.05$). The prevalence was significantly greater in rural schools (4.8/1000) than in urban schools (1.98/1000) ($p<0.05$). There was overcrowding and poor housing in most cases. There were fewer cases of RHD with severe valvular lesions in the younger age group than in the older children. The mitral valve was the valve most commonly affected by RF/RHD.

Conclusions – RHD continues to be a serious health problem. Regular surveys are needed to identify cases early and to ensure secondary prophylaxis with penicillin is given thereby preventing recurrence of RF and progression of the severity of the valvular lesion. Echocardiography is necessary to identify cases of RF/RHD. Strategies for preventing RHD should involve primary prevention to avert the first attack of carditis and strengthening of secondary prophylaxis through improved education and motivation of patients, parents, and physicians.

(RF)/RHD and this constitutes a major health problem.¹ The extent and nature of the problem has been well documented by both hospital and community based studies. This malady affects adversely a large proportion of the population. It has been estimated that about two million children are affected by this disease in India. A multicentre study on the epidemiology of RHD, under the aegis of Indian Council of Medical Research (ICMR) gave the national average of RHD prevalence as 6 per thousand in the age group of 5–16 years.² The only preliminary study undertaken in the hilly area of Himachal Pradesh was in 1956. This showed a prevalence of 39 per thousand among the 1515 school children in Shimla, the highest prevalence ever reported in India.³ The obvious pitfall of this study was that investigating facilities were meagre then and there was considerable lack of awareness among the public about this illness. We therefore undertook a school based study to investigate the current extent of the problem.

Methods

GEOGRAPHIC AREA

Himachal Pradesh is a picturesque hilly state in northern India, with a population of 5 170 877 and a population density of 93 km². The study was based on a survey carried out among school children in Shimla town and the adjoining area of Kasumpti-Suni Block, Shimla District, Himachal Pradesh.

METHOD OF SAMPLING

All the schools in Shimla town and Kasumpti-Suni Block were grouped into four categories – rural government, urban government, convents, and private schools. There were 141 rural government, 99 urban government, 15 convent, and 9 private schools in the survey area, and of these 31, 18, 8, and 5 respectively were selected randomly. A stratified random sample of 16 082 children out of 40 950 children in the age group of 5–16 years from these schools were selected for the study.

All the children present in a class at the time of the visit were examined in one sitting by a doctor (JST) who had trained for six months in cardiology especially for this purpose. History of past and present joint pains, fever, sore throat, palpitation, fatigue, shortness of breath, and chest pain were elicited and recorded on an initial screening proforma. Clinical examination

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Rheumatic heart disease (RHD) is a problem that affects each corner of the globe, especially the developing countries. In India, 32–50% of all cardiac cases result from rheumatic fever

Table 1 Age and sex distribution of children screened

Age group (y)	Children screened		Children recalled		Suspected RF/RHD		Definite RF/RHD	
	M	F	M	F	M	F	M	F
5–10	3567	2987	262	237	16	12	7	8
11–16	4553	3973	292	302	28	45	16	14

M=male; F=female; RF=rheumatic fever; RHD=rheumatic heart disease

was performed to look for objective evidence of RF and RHD. Particular care was taken in auscultating the child in erect, left lateral, and recumbent positions. In doubtful cases, children were also auscultated after exercise. An attempt was made to include the absentees during subsequent visits or by home visits (maximum two visits).

Parents of the children aged below 10 years and all the children aged above 10 years were asked about the family income. A modified Prasad’s classification based on family income was used to determine the socioeconomic status of the children.⁴

The following types of patients were then recalled to the Indira Gandhi Hospital to be examined by a cardiologist to rule out observer’s bias:

- Suspected and definite cases of RHD;
- History of RF with or without carditis;
- History suggestive of RF with or without carditis;
- Past history of RF or previous history suggestive of RF;
- Suspected and definite cases of congenital heart diseases;
- Patient with an ejection systolic murmur with grade I–II/VI functional murmur with or without history of RF.

The following types of patients were subjected to detailed echocardiographic examination after evaluation by the cardiologist:

- Suspected and definite cases of RHD and congenital heart disease;
- History of RF with or without carditis;
- Past history of RF;
- Previous history suggestive of RF with functional murmur.

These cases were also subjected to following investigations: chest x ray (PA view); 12 lead electrocardiogram, haemoglobin, total leucocyte count, differential leucocyte count, erythrocyte sedimentation rate, ASO titre, and C reactive protein; and a throat swab for β -haemolytic streptococci (BHS). An Advanced Technology Laboratories (ATL) Ultramark 7 echocardiograph with facilities for 2D, M mode, and Doppler ultrasonography was used for the study. Jones revised criteria were used for the diagnosis of RF.⁵

A home visit was made to the diagnosed cases of RF/RHD to study the housing conditions in accordance with WHO expert committee criteria for healthful housing and also the personal observation of examiner.

For analysis, the following definitions were used:

- **Acute RF:** those patients who fulfilled revised Jones criteria at the time of screening, with no history of RF in the past.
- **Probable RHD:** thickening of mitral and/or tricuspid valve seen on echocardiography, with previous history of RF or thickening of aortic valve and without Doppler evidence of valvular dysfunction.
- **Definite RHD:** clinical and echocardiographic evidence of chronic RHD.

Results

AGE AND SEX DISTRIBUTION OF THE CHILDREN

The age and sex distribution of the children is shown in Table 1. A total of 15 080 (93.7%) school children were examined out of a sample of 16 082 children selected for the study. Altogether 8120 (53.8%) of the total children screened were boys whereas only 6960 (46.2%) were girls. 6554 (43.5%) children were in the age group of 5–10 years and 8526 (56.6%) were in the age group of 11–16 years. There was almost equal distribution of male and female children in both age groups ($p>0.05$).

PREVALENCE OF RF AND RHD

Out of 15 080 children screened, 1116 (7.4%) with a history of RF and a heart murmur on clinical examination, or both, were called for detailed examination by a cardiologist. Twenty three (2%) children were non-responders – nine were suspected RF/RHD and congenital heart disease (CHD), and five had a functional murmur.

Suspected and definite cases of RHD

After a second screening by a cardiologist, 101 cases were suspected of having RF/RHD (table 1) and 49 were suspected of having CHD. The rest of the children were found to have a functional murmur or a history of non-rheumatic joint pains and hence did not undergo further investigations. All the children suspected of having RF/RHD and CHD had echocardiography and other investigations. A past history of RF with or without carditis was present in 38 (37.6%). Fifty seven (56.4%) had a history suggestive of RF, with or without carditis, and in six (5.9%) there was no past history of RF.

A clinically definite diagnosis of RHD was made in 26 (25.7%) children, confirmed on echocardiography, and a probable diagnosis in 75 (74.25%) cases. Six (8%) of the clinically probable group were found to be suffering from definite RHD and 12 (16%) had echocardiographic evidence suggestive of RHD. Of the six with definite RHD, three (50%) had Gr II/VI ejection systolic murmur at the mitral area and three (50%) had no murmur but a past history of RF. In total, therefore, there were 32 (31.68%) cases of definite RHD and 12 (11.88%) of probable RHD on echocardiography, giving the overall prevalence rate of RHD as 2.92 per thousand.

There were three clinically definite cases of acute RF, two had carditis confirmed on echocardiography but the third had no carditis and

Table 2 Prevalence of rheumatic heart disease (RHD) and the incidence of acute rheumatic fever (RF) and rheumatic fever recurrence in different types of schools

Type of school	Definite RHD	Probable RHD	Overall prevalence	Acute RF	RF recurrence
Rural government (n = 4989)	19 (3.8)	5 (1)	24 (4.8)	1 (0.2)	3 (0.6)
Urban government (n = 3884)	9 (2.32)	1 (0.26)	10 (2.57)	1 (0.26)	1 (0.26)
Convent (n = 4359)	2 (0.46)	5 (1.15)	7 (1.6)	0	0
Private (n = 1848)	2 (1.08)	1 (0.54)	3 (1.62)	1 (0.54)	0
Total n = 15080	32 (2.12)	12 (0.8)	44 (2.92)	3 (0.19)	4 (0.26)
	$\chi^2 = 13.40$; (p<0.01)	NS	$\chi^2 = 9.94$; (p<0.05)	NS	NS

Figures given in parentheses are the prevalence/incidence rate per thousand

Table 3 Distribution of rheumatic fever/rheumatic heart disease (RF/RHD) cases according to socioeconomic status

Social class	Total children (%)	No of RF/RHD cases	RF/RHD prevalence per thousand
I	1756 (11.6)	0	0
II	2812 (18.6)	7	2.49
III	4052 (26.9)	11	2.71
IV	4280 (28.4)	17	3.97
V	2180 (14.5)	10	4.59
Total	15080 (100)	45	2.98

$\chi^2 = 8.87$; (p>0.05)

echocardiography was normal. This gave an incidence rate for acute RF as 0.19 per thousand. There were four cases with RF recurrence. This was confirmed by laboratory investigations and echocardiography, giving an incidence rate of 0.32 per thousand. Two patients with acute RF had developed RHD with valvular dysfunction at the time of study and hence were included among 44 RHD cases. In total there were 45 cases of RF/RHD out of 15 080 school children, giving an overall prevalence of 2.98 per thousand. Thus, out of 1093 children selected for detailed examination/investigations, 45 had RF/RHD, 34 had CHD, 912 had a functional murmur, and 102 had non-rheumatic joint pains. The prevalence of functional murmur was 6.04%.

Initial screening and subsequent visit to screen absentees
During the initial screening 13 020 children were examined, out of whom 42 had RF/RHD. On subsequent visits to screen children who had been absent at the first visit, three cases of RF/RHD were found in 2060 children, giving prevalence rates of 3.22 and 1.45 per thousand respectively.

AGE AND SEX DISTRIBUTION OF RF/RHD CASES
The mean (SD) age of the children with RF/RHD was 12.04 (3.1) years, ranging from 6 to 16 years. There were 30 cases (3.5/1000) of RF/RHD in children aged 11–16 compared with 15 cases (2.29/1000) in the 5–10 years group as shown in table 1. This difference was not statistically significant (p>0.05). There were 23 boys (2.83/1000) and 22 girls (3.16/1000) with RF/RHD but this difference was not significant (p>0.05).

PREVALENCE OF RHD IN DIFFERENT CATEGORIES OF SCHOOLS
This is shown in table 2. The prevalence of definite RHD and the overall RHD prevalence

was significantly higher in rural government schools (4.8/1000) and lowest in convent schools (1.6/1000) (p<0.05).

PREVALENCE OF RHD IN RURAL AND URBAN AREA
The prevalence of RHD was significantly higher in rural (4.8/1000) than in urban areas (1.98/1000) (p<0.05). The prevalence of definite RHD was also significantly higher in rural (3.81/1000) than in urban areas (1.29/1000) (p<0.05).

SOCIOECONOMIC STATUS
All the children screened were grouped into five social classes as given in table 3. The prevalence of RF/RHD was high among lower social classes, but these differences were not significant (p>0.05).

HOUSING
In most cases, that is 33 (73.3%), housing conditions were poor, there was no proper environmental sanitation and considerable overcrowding. Only 12 (26.7%) lived in good conditions.

PRESENTATION OF PATIENTS WITH RHD
Most of the cases presented with a history of recurrent joint pains (84%), and migratory joint pains were present in 21 (47.7%). A current history of joint pains was present in 10 (22.72%) cases, out of whom eight (18.2%) had arthralgia and two (4.5%) had arthritis. Of two cases of arthritis, all the major joints were involved in one and the other had knee joint involvement. Twelve (27.2%) cases presented with a history of an antecedent sore throat. A history of fever at the time of screening was present in five (11.36%) cases and 22 (50%) cases presented with recurrent fever. Breathlessness and palpitation were present in 34 (77.3%) and 15 (34.1%) cases respectively. Chest pain was present in seven (15.9%) cases. The New York Heart Association (NYHA) functional classes of the patients were as follows: class I = 13 (38.2%); II = 13 (38.2%); III = 7 (20.6%); IV = 1 (2.9%).

CLINICAL AND ECHOCARDIOGRAPHIC PROFILE OF PATIENTS WITH RHD
This is shown in table 4. Of 44 cases of RHD, only 29 (65.9%) gave a past history of RF, nine (20.5%) gave a previous history suggestive of RF, and in six (13.63%) there was no previous history of RF. Two cases who gave a past history of RF had only Gr II/VI functional murmur at the clinical examination but they were found to have mild mitral regurgitation on echocardiography. Similarly, one child with a previous history suggestive of RF and grade II/VI functional murmur was found to have definite RHD.
Of 44 cases, 10 (22.7%) were asymptomatic and 34 (77.3%) were symptomatic. On clinical examination, there was murmur in 29 (65.9%) cases. Three (6.8%) cases who were normal on

Table 4 Clinical and echocardiographic profile of patients with rheumatic heart disease (RHD)

Rheumatic fever status	Clinical CVS findings	Echocardiographic findings	No of patients
Past history of RF (n = 29)	Definite RHD (n = 17)	Definite RHD	17
	Gr II/VI ESM at apex (n = 2)	Definite RHD (mild MR)	2
	Normal (n = 3) (one case was with chorea)	Definite RHD (mild MR)	3
	Normal (with chorea) (n = 1)	Probable MV disease	1
	Normal (n = 5)	Probable MV disease	5
	Gr II/VI ESM at apex (n = 1)	Probable MV disease	1
Previous history suggestive of RF (n = 9)	Definite RHD (n = 3)	Definite RHD	3
	Gr II/VI ESM at apex (n = 1)	Definite RHD (mild MR)	1
	Normal (n = 4)	Probable MV disease	3
		Probable MV and AV disease	1
No past history of RF (n = 6)	Gr II/VI ESM at apex (n = 1)	Probable MV disease	1
	Definite RHD (n = 6)	Definite RHD	6
Total			44

MV = mitral valve; AV = Aortic valve; MR = Mitral regurgitation; CVS = cardiovascular system; ESM = ejection systolic murmur

Table 5 Type of valvular lesion

Type of lesion	No of children			% of total	
	Male	Female	Total	Male	Female
MR	6 (40.0)	10 (58.2)	16 (50.0)	37.5	62.5
MS	1 (6.7)	0	1 (3.1)	100	0
AR	1 (6.7)	0	1 (3.1)	100	0
MR + MS	1 (6.7)	1 (5.8)	2 (6.3)	50	50
MR + AR	2 (13.3)	2 (11.8)	4 (12.5)	50	50
MR + TR	2 (13.3)	2 (11.8)	4 (12.5)	50	50
MS + TR	1 (6.7)	1 (5.8)	2 (6.3)	50	50
MS + AR + TR + TS	0	1 (5.8)	1 (3.1)	0	100
MR + MS + TR + TS + AR + AS	1 (6.7)	0	1 (3.1)	100	0
Total	15 (100)	17 (100)	32 (100)		

Figures given in parentheses indicate percentage
MR = mitral regurgitation; AR = aortic regurgitation; TR = tricuspid regurgitation; MS = mitral stenosis; AS = aortic stenosis; TS = tricuspid stenosis.

clinical examination were found to have definite RHD on echocardiographic examination. A previous history of chorea was present in five (11.4%) cases. No case gave past history of subcutaneous nodules or erythema marginatum. Pharyngitis was present in 24 (54.5%) cases at the time of examination. Two (4.54%) cases had congestive cardiac failure. Pulmonary arterial hypertension was present in nine (20.5%) cases and pulmonary venous hypertension in five (11.36%) cases. Endocarditis was present in one (2.3%) case and pedal oedema in two (2.3%).

VALVULAR INVOLVEMENT AND TYPE OF VALVULAR LESION
Of the 44 cases, the mitral valve was involved in 43 (97.7%), the aortic valve in nine (20.5%), and the tricuspid valve in eight (20.5%). No case of pulmonary valve involvement was found. The type of valvular lesion is given in table 5. Of the 44 cases of RHD, 32 (72.7%) had definite RHD and the remaining 12 (27.3%) had echocardiographic evidence of valvular involvement with no definite valvular dysfunction. Mitral regurgitation was the commonest lesion (50%) in these 32 cases.

SEVERITY OF VALVULAR DISEASE IN RHD CASES
One of 15 cases (6.7%) in the 5–10 years group had a severe valvular lesion compared with five of 29 (17.2%) in 11–16 years age group.

LABORATORY INVESTIGATION IN RHD CASES
The ASO titre was raised in six (13.6%) cases; the C reactive protein test was positive in only one (2.3%) case; the erythrocyte sedimentation rate was raised in 11 (25%) cases; and throat swab culture for β haemolytic streptococci (BHS) was positive in 20 (45.5%) cases. Five cases (11.4%) had both a raised ASO titre and throat swab positive for BHS. Clinical examination showed that pharyngitis was present in 24 (54.5%) cases, 14 (58.2%) of whom were positive for BHS.

SECONDARY PROPHYLAXIS
Only 27 of the 44 (61.4%) cases were taking secondary prophylaxis with penicillin at the time of the study. Of these, 10 (37%) who were on secondary prophylaxis were positive for BHS on throat culture and only two were taking it irregularly. Of four cases with RF recurrence, only two (50%) were on secondary prophylaxis, though they were taking it irregularly.

PROFILE OF THE FIRST ATTACK OF RF
Out of the three cases of acute RF, two (66.7%) had carditis and one (33.3%) had no carditis. Arthritis was seen in two (66.7%) cases. Subcutaneous nodules were present in one case only. No case was seen with chorea and erythema marginatum. Two cases of acute RF were positive for BHS on throat culture.

Discussion
This study tried to determine the prevalence of RHD/RF in school children aged 5–16 years in the state of Himachal Pradesh. This work is the first of its kind in the last 30 years and covers a large number (n = 15 080) of school children. It is one of the largest studies undertaken in India.
In this study, all 26 cases found to be clinically definite were confirmed by echocardiography. However, only 24% of clinically probable cases were found to have RHD by echocardiography, thereby indicating the low specificity of clinical observation. Hence, identification of RHD by clinical criteria is bound to overdiagnose the disease in community surveys. Similarly, six children whose diagnosis was uncertain after clinical examination were found to be suffering from definite RHD echocardiographically, indicating a low sensitivity of the clinical diagnosis. Echocardiography also enabled us to make a probable diagnosis of RHD in 12 (16%) of 75 clinically suspected RHD cases in whom there was echocardiographic evidence suggestive of RHD. The roles of echocardiography and Doppler ultrasonography as highly sensitive and specific tools in the assessment of RHD have been emphasised in various studies from all over the world.^{5,6}
In our target population (5–16 years) of 15 080, the prevalence of RF/RHD was 2.98 per

thousand, comparable with a recently published report of the prevalence of RHD (2.2 per thousand) in 16 developing countries of five WHO regions.⁷ The prevalence of RF/RHD in the present study was less than the 11 per thousand reported from Sahafa town, Sudan,⁸ comparable with 2.4 per thousand reported from Saudi Arabia⁹ but higher than the 1.7 per thousand reported from western Kenya¹⁰ and is also slightly higher than that reported from our neighbouring countries.^{11,12}

If the prevalence rates of RHD in different parts of India are compared our rate was less than those of Calcutta and Hyderabad² and slightly higher than those of Alleppy, Bombay² and Anand. In northern India, the prevalence of RHD in our study was comparable with those in neighbouring states¹³⁻¹⁶ but lower than that in Delhi.¹⁷ It was observed that there was not much difference in the prevalence of RHD where echocardiography had been used.¹⁴⁻¹⁶ The prevalence of RHD in our study was higher in rural areas (4.8/1000) a finding also reported from Saudi Arabia⁹ and from school children in a country town in Ethiopia.¹⁸ Koshi *et al* reported a RHD prevalence of 4.9 per thousand in rural south Indian children.¹⁹ In a recent survey from rural area of Rajasthan, the prevalence of RHD was 3.34 per thousand where echocardiography had been used.¹⁵ So the problem of RHD is still higher in rural areas of India.

The RHD prevalence was highest in government schools in our study as in other studies.^{17,20} It was slightly higher in the 11-16 years age group than in the 5-10 years groups, in agreement with other studies.^{14,17} The RHD prevalence was almost equal in both sexes, as has been reported by others.^{10,11,16,17} The prevalence of RF/RHD on subsequent visits to screen those children who were absent at the first visit was lower (1.45/1000) than in the initial screening (3.22/1000). This may be due to some other causes than RF/RHD but every effort should be made to achieve as high a coverage as possible to get true picture of the disease.

There was a clear trend of an increasing prevalence of RHD with a fall in the socioeconomic status, though the difference was not significant as in other studies.¹³⁻¹⁶ It has been reported that the prevalence of RF/RHD increases directly with overcrowding and poor housing.⁸ Overcrowding and poor housing were present in most of our subjects though no valid inference could be drawn due to lack of controls for comparison.

About 66% of our cases gave a past history of RF, compared with 62% in the study of Berry¹² and 40% in a study of Shimla by Chand.¹¹ Eighty four per cent of the children with RHD gave past history of recurrent joint pains, 48% of migratory joint pains, and 11.36% of chorea hence chorea was not as uncommon as has been believed by others.

In a school survey in Delhi, 18% of cases of RHD gave past history of polyarthritis, 26% of migrating polyarthralgia, and 3% of chorea.¹⁷ Berry found polyarthritis, polyarthralgia, and fever as the commonest combination and a history of chorea was present in only six cases.²¹ Subcutaneous nodules are also not as uncommon as previously thought. Erythema mar-

ginatum has been reported from India by many workers but was not seen here.²¹

Isolated mitral regurgitation was commonest lesion, as had been reported in India by many workers.^{13,17,20} It was followed by combined mitral and atrial regurgitation and combined mitral and tricuspid regurgitation, each found in 12.5% of cases. Isolated mitral stenosis and isolated atrial regurgitation were present in 3.1% cases each. Isolated mitral stenosis in the present study was less frequent than reported by others.^{17,20}

RHD with severe valvular lesion was more common in the 11-16 years age group than in the children aged 5-10 years. There is therefore a need to identify cases early when the valvular lesion is less severe and to give them secondary prophylaxis with penicillin thereby preventing recurrence and progression of the severity of the valvular lesion.

About a quarter of the cases of RHD were asymptomatic: these children did not attend the hospital and were not taking secondary prophylaxis with penicillin. Survey studies are therefore needed to identify these asymptomatic patients.

The carrier rate of BHS had been reported as 34% in Saudi Arabia⁹ and 24% in RHD cases in Delhi.¹⁷ Our carrier rate was 45%, which can be attributed to the poor living conditions prevailing in the area. Despite secondary prophylaxis in 27 cases, 10 (37%) children were positive for BHS on throat culture and only two of them were taking penicillin irregularly.

Some 38.6% of RHD cases were not taking secondary prophylaxis with penicillin. The role of secondary prophylaxis in RHD patients has been emphasised by many studies.⁵ These patients need to be educated about the importance of secondary prophylaxis.

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