Epidemiology and Prevention of Rheumatic Fever and Rheumatic Heart Disease

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Rheumatic fever and rheumatic heart disease still persist as one of the main health problems in developing countries. It is a primary cause of hospitalization for heart diseases, constituting 1/4-1/2 of children. While the disease is on the decline in many parts of the world, it is so tragic that in our country we still watch helplessly many children and young adults dying of advanced rheumatic heart disease.

INCIDENCE AND PREVALENCE

Our knowledge of incidence and prevalence of rheumatic fever is rather limited as it is extremely difficult to obtain reliable data specially from developing countries where rheumatic fever is much greater problem. Many studies do not state the denominator of population¹, nor the criteria used for diagnosis and hospital admissions and, therefore, do not give a true picture of the occurrence of the disease.

There are not many reported surveys from our country but Ilyas, M. et al² conducted separate studies on the pattern of rheumatic fever in the period of 1966-79 which showed a preponderance of cardiac lesion (65% - 78%) over arthritis (30%- 59%) and skin manifestations (2.5-10%). Rhumatic heart disease surveys in school children have shown a low prevalence (91.8 per 1000) in selected high income group in Karachi, a high prevalence (7 per 1000) in Peshawar and still higher (11 per 1000) in Chitral. Hospital population data show rheumatic heart disease as the most common cardiovascular disorder in children and adoelscents constituting 4% of all medical cases and 23% of all cardiac cases³.

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DECLINE OF RHUMATIC FEVER

It has become a fact that both the incidence of rhumatic fever and prevalence of rhumatic heart disease are declining in developed countries. The best information comes from Denmark where rhumatic fever (R.F.) has become a notifiable disease since 1978. In Japan the prevalence of rhumatic heart disease in school children, which was 4.6 per 1000 in 1958, has decreased spectacularly to 0.1 per 1000 by 1971. Similar figures are obtained from most of the developed countries⁴.

The reasons for the decline are undoubtedly multiple and are listed below:

1) Improved standard of living which means better housing, less crowding and improved nutrition.

2) Greater access to improved health care.

3) Wide-spread use of antibiotics.

4) Decline in streptococcal infection.

5) Diminished streptococcal virulence.

6) Fever "rheumatogenic" serotypes.

The severity of the disease also seems to be getting less in developed countries as studies from Europe and Japan show less cases of pure mitral stenosis and more of mild mitral regurgitation⁵.

The pattern of mortality also seems to have changed in these countries as in U.K. in 1940 most deaths in rheumatic heart disease were due to heart failure; whereas in 1980, they are mostly due to cardiac surgery.

In developing countries the picture is quite different as the disease is much more aggressive because of the severity and early onset of the disease;
one wonders if it runs a different clinical course as compared to the one in developed countries.

However, there are some good studies from India\textsuperscript{6,7} which suggest that the relationship of R.F. to the intensity and severity of streptococcal disease is the same in tropics as in the temperate climates. The clinical features of rheumatic fever are also alleged to be different in different climates, being more cases of carditis in tropical climates\textsuperscript{5}.

A study from India showed that more than half of 8000 patients had isolated mitral stenosis and of these more than 1/4 were below the age of 20 (Juvenile M.S.). More than 75\% of these had severe symptomatic disease.

A good perspective study of clinical profile of rheumatic fever in India by Sanyal et al\textsuperscript{7} has emphasised that the first attack of rheumatic fever in children closely resembles the clinical picture in developed countries.

The difference occurs only with recurrent episodes of rheumatic fever which lead to worsening of the disease.

The difference between the two presentations may be due to overwhelming socio-economic disparity confirming the old saying that ‘rheumatic fever is a social disease affecting the poor’.

The early onset of rheumatic fever in our community may be due to large number of children in the family with greater exposure to streptococcal infection because of poor living conditions. Similarly the severity may be due to higher rate of recurrence with repeated greater exposure and lack of medical care.

**STREPTOCOCCAL PHARYNGITIS AND ACUTE R.F.**

Acute R.F. develops two weeks after group A beta haemolytic streptococcal throat infection as some type of hyper-immune reaction due either to bacterial allergy or auto-immunity.
The bacteria contains surface M protein antigen of which nearly 80 serotypes have been identified up till now. The bacterial cell wall contains peptides, polysaccharide-complexes which persist for several months in macrophages and later on occur in granulomas in heart, synovium and liver.

The streptococci also produce several extracellular toxins, streptolysin S&O, protein-ases, streptokinase, deoxy-ribo-nuclease, erythrogenic & cardio-hepatic toxins. Antibodies to these toxins, produced by the host, allow identification of recent streptococcal infection.

**IMMUNO-GENETIC SUSCEPTIBILITY**

No discussion of R.F. is complete without mentioning genetic predisposition to R.F.

Since acute R.F. develops in only a small percentage of patients following even the most virulent strep. infection, one wonders if host genetic predisposition plays a strong role. It has been found that individuals with certain types of HLAB alloantigen, in particular 833, 1818, are more susceptible to R.F.

**PREVENTION**

R.F. is a preventable disease if infections with group A streptococcal sore throat are diagnosed early and treated adequately and promptly.

To control R.F. one has to understand its epidemiology, the methods available locally to prevent its recurrence and the most important to consider local socio-economic set up of a particular country.

Like many developing countries, in Pakistan there are two extremes of social classes: on the one hand there is unlimited wealth, culture and sophisticated technology and on the other hand there is extreme poverty, primitiveness, ignorance and illiteracy. Whatever resources are available are misused or under-utilised. Therefore, the approach has to be realistic to be successful. Realistic goals are as follows:
1) Improvement in social economic condition which means better housing and less crowding, adequate financial support to all citizens to use good diagnostic and therapeutic management of streptococcal infections and eradicating ignorance and illiteracy.

2) Immunisation against group A beta haemolytic strept. has not become a reality until now because of difficulty of many serotypes of surface M-protein antigen. There is lot of work going on its purification and it is hoped that there will soon be a much improved purified polyvalent type specific M-protein vaccine for clinical use which will be a break through in the control of R.F. and streptococcal infection.

3) Chemoprophylaxis can be divided into:
   i) Primary prophylaxis: prevention of initial rheumatic attack.

CHEMOPROPHYLACTIC PROGRAMME WITH BENZATHINE PENICILLIN

1) Primary prophylaxis:
   a) 1.2 mega units IM single dose per patient, age 18 years or over.
   b) 0.6 mega units IM single dose per patient, age less than 18 years.

2) Secondary prophylaxis:
   a) 1.2 mega units IM every 3 weeks for patient age 18 years or over.
   b) 1.2 mega units IM every 4 weeks for patient age less than 18 years.
PRIMARY PREVENTION

Classic work on army recruits in USA clearly showed that in epidemic conditions, the attack rate decreased from 2.8% in untreated group to 0.2% in treated group.8

Group A haemolytic strept is still very sensitive to pencillin. The most effective way of eradicating streptococcal throat infection is by a single dose of repository form of long-acting benzathine pencillin which has proved to be the drug of choice. It is least expensive, least allerginic of all pencillins with less possibility of anaphylaxis. It has the advantage of single injection with more definitive effective tissue levels.

The fear of anaphylaxis is highly exaggerated and ill-founded. It is seen more commonly in adults than children. It must be mentioned here that skin test has no value whatsoever in predicting which patient will develop anaphylaxis.

Oral pencillin (pencillin V) 250 mg BD is more expensive with risk of poor compliance and unsatisfactory blood levels.

Patient allergic to pencillin can be given erythromycin 250 mg BD or sulphadiazine IG OD over 60lbs of weight and 1/2 G BD under 60lbs of weight. There have been recent reports of resistant strains to sulpha. The injection should be given to high risk children during peak streptococcal infection (wet seasons) or epidemics of sore throat. Places with overcrowding like schools, hostels and barracks should be carefully observed. The families of rheumatic fever cases should also be kept under observation and treated properly if and when necessary. The carriers of streptococcal throat infection should be identified and treated.

PROBLEMS OF PRIMARY PROPHYLAXIS

1) Under diagnosis of streptococcal infection.
2) Large number of subclinical cases with no symptoms or history of sore throat.
3) Inadequate facilities for proper culture.
4) Inappropriate therapy.
5) Since only 0.3% - 3% of strepto sore throat will develop acute R.F., a large population has to be given injections un-necessarily with tremendous burden on the national budget.

Because of the above problems primary prophylaxis cannot prove to be effective in our country, therefore, all the efforts and energy should be used in stressing the importance of secondary prophylaxis.

SECONDARY PROPHYLAXIS

The more realistic goal of high priority is prevention of recurrence of rheumatic fever.

FACTORS WHICH INFLUENCE THE RECURRENCE OF R.F.

1) Severity of streptococcal pharyngitis.
2) Presence of existing R.H.D.
3) Number of preceding rheumatic attacks.
4) Recency of last attack.

A) Rheumatic subjects become supersensitised after initial attack of streptococcal infection so that the recurrence can be as high as 65% as compared with 3% of non-rheumatic subjects.

B) Carditis occurs in 64% - 80% of patients with acute R.F. in developing countries.

C) Presence of carditis with initial R.F. will result with more carditis with subsequent attacks leading to severe rheumatic heart disease.

D) Carditis with initial attack may resolve in 9.4% to 80% of patients provided it is mild and recurrence of R.F. is adequately prevented. Benzathine penicillin is the drug of choice with low recurrence rate as compared to oral route. A supervised subsidised community based programme in Phillipines has shown a decrease from 4.4% to 0.5% of recurrence of R.F.
Length of prophylaxis is in little dispute. While it is generally accepted to be five years or till the age of 20, whichever comes first. WHO experts recommend life long prophylaxis for developing countries.

Patients after valve surgery are in greater need of life long prophylaxis.

Some studies have shown 3 weekly injections to be more effective than 4 weekly in developing countries.

Patients allergic to penicillin can have erythromycin. Sulphonamide should not be used because of resistance.

Tonsillectomy has no place in secondary prophylaxis as it will not protect patient to have recurrence.

It must be remembered that secondary prophylaxis for R.F. will not eliminate the risk of bacterial endocarditis. Therefore, patients on long acting penicillin should receive additional antibiotics for transient bacteraemias to cover the risk of S.B.E. The main problem of secondary prophylaxis is non-compliance which may not at times be due to patient's fault. There may be non-availability of penicillin or the staff for injection. Lack of education, ignorance, remoteness of the country side and poverty may be contributing factors in non-compliance.

**FAILURE OF CONTROL OF R.F. IN PAKISTAN**

1) Unsatisfactory improvement in socio-economic conditions.
2) Lack of concentrated effort for its prevention.
3) Ignorance of epidemiological lesson that the disease is preventable.
4) Uncertain approach towards primary prophylaxis with no proper school health programme.
5) Inadequate secondary prophylaxis with no subsidised supervised community based programme.

In summary we must start a realistic approach of control of R.F. as soon as possible with properly supervised and subsidised community based
programmes as sponsored by WHO. These programmes have been started in several developing countries with initial good results.

These programmes should involve all concerned government agencies, private voluntary charity organisations, hospitals, clinics, schools, doctors, nurses auxiliaries etc. A R.F. register should be kept with proper compliance surveillance teams.

The cost benefit ratio of this programme would be clear when one knows that the staggering cost of one mitral value replacement would be sufficient for 5 years prophylaxis of 100 children.

REFERENCES


