

Review Articles

PREVENTION OF RHEUMATIC FEVER

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Rheumatic fever is unique among the major causes of cardiovascular diseases because it is the only one that is definitely preventable. Nevertheless, it still occurs with unacceptable frequency. Effective methods for prevention of first attacks and recurrences of RF are available, but are not being applied to the best advantage of all populations enjoying near optimal medical care, this is not true for populations in the developing world¹.

Rheumatic fever (RF) and rheumatic heart disease (RHD) constitutes major health problems today in the developing countries because of their great impact on the afflicted individual and his community. Further successful control and prevention of these diseases require a multifactorial approach by many medical and paramedical personnel in addition to the practising physician. The skill of the state and the local health officers, social workers, health educators and laboratory technicians are all necessary. Identifying rheumatic patients and maintaining them on longterm prophylaxis against streptococcal infections depend on a co-operative community-wide program of official, voluntary and private agencies as well as medical institutions and physicians in practise².

The most effective measures against RF are probably socioeconomic. The almost total absence of the RF from affluent sections of the cities of the Western World suggests that spacious housing and non-crowding are at least as important as good diagnosis and treatment of streptococcal infections³.

Prevention of RF is considered in three levels, first prevention of the occurrence of the disease, this is the primary prevention; second prevention of recurrence of the disease after the first attack, this is the secondary prevention; lastly prevention of further disability and death, and this is the tertiary prevention⁴.

PRIMARY PREVENTION

Primary prevention consists of identification and diagnosis of upper respiratory streptococcal infection, and proper treatment with proper antibiotics in a proper dose for the proper time.

DIAGNOSIS OF STREPTOCOCCAL INFECTIONS

CLINICAL DIAGNOSIS:

One of the obstacles of prevention of initial attacks of RF is the difficulty of clinical diagnosis of streptococcal nasopharyngitis. The sudden onset of fever, headache, dysphagia, beefy redness of the pharynx with exudate, nausea and vomiting may occur specially in children, scarletiform rash, and tender cervical nodes represent characteristic syndrome of streptococcal sore throat. A scarlatinal rash is diagnostic when it occurs but it is much less frequent than formerly. Scarlet fever is also less severe and at times the rash is less typical and difficult to distinguish from other exanthema. In children over two years of age, otitis media is often caused by group A streptococci, and such patients should receive recommended therapy. The classic findings of streptococcal disease are more likely to be present during epidemics but occur only occasionally in patients with sporadic infections. The diagnosis is more difficult when the tonsils are absent or when the patient is seen only once during the course of illness, a fairly common occurrence at present time.

Several studies have shown that the physician can diagnose streptococcal infection clinically in 55-70% of the cases. The chief problem is to distinguish mild streptococcal illness from viral infections of upper respiratory tract. Conjunctivitis, coryza, hoarseness and tracheitis are more likely to be a viral infection. However, it is not possible to distinguish bacterial from non-bacterial disease on the basis of pharyngeal redness alone. Indeed, even the occurrence of exudate, often considered the hall mark of streptococcal infection, may not always be a reliable sign. It has been described with adeno and Coxsackie virus infection.

LABORATORY AIDS:

To differentiate between viral and mild streptococcal

infection W.B.C. and throat cultures are useful tools. A W.B.C. of more than 12000 cells indicates that the infection is most likely to be bacterial. A sure way of differentiation is to grow the streptococci in the culture; throat swab inoculated in blood agar showing more than 10 colonies indicates that streptococci play an etiologic role. In many patients with clinical symptoms, typing of streptococci is unnecessary because it is mostly due to group A haemolytic, whereas in special circumstances identification of group A haemolytic by bacitracin disc is needed. A rapid method of such identification is by the use of immunofluorescent antibody staining techniques.

Antistreptolysin O (ASO) or other streptococcal antibody test (Anti DNase, antihyaluronidase, antistrep tokinase, ant NADase) are useful in confirming a recent group A streptococcal infection and are therefore helpful in patients presenting with possible non-suppurative complication of streptococcal infection. However, they are of no immediate value in the diagnosis and management of acute streptococcal infection⁵.

Miscellaneous streptococcal infection :

Infections of the skin and vagina are not uncommon. In 40% of patients with impetigo evidence of streptococcal infection can be shown. Initial or recurrent attacks of RF, unlike AGN, rarely if ever follows skin streptococcal impetigo because these lesions are superficial and evolve little immune response⁶.

Treatment of Streptococcal Infections

Treatment should begin immediately in ill, toxic children with symptoms and signs highly suggestive of streptococcal pharyngitis. Treatment should be commenced after the result of throat cultures in cases of less characteristic findings. Penicillin therapy is still effective despite the delay due to culture. Once the diagnosis is clinched the patient should be treated no matter how mild the clinical symptoms. The aim is to eradicate streptococci from the nasopharynx. Mere clinical recovery is not alone sufficient. Persistence of organism is common following inadequate treatment

and this is the chief reason for failure to prevent RF.

The properly selected antibiotic must be administered in sufficient dosage to maintain therapeutic blood levels for ten days. Sulphonamides should not be used for treatment of streptococcal infections, also tetracyclines because of increasingly development of resistant strains^{5,7}. Penicillin is the drug of choice except in patients allergic to it. Fortunately, group A streptococci have remained exquisitely sensitive to penicillin. The most effective procedure is a single 1/M injection of benzathine penicillin, 600 000 units for less than 10 year olds, 900 000 units for more than 10 year olds, and 1.2 megaunits for adults. It is effective in 95% of patients. Single daily injections of aqueous or procaine penicillin is inadequate and generally impracticable.

Oral penicillin V, 200-250 000 units 3 or 4 times daily for 10 days may be used. The chief disadvantage is the failure to take the drug regularly for 10 days. Bacterial and clinical relapses occur in 15% of patients. Usually a second course in a higher dose will eradicate the organisms. Treatment failure is due to penicillinase-producing Staphylococci strains, addition of antistaphylococci antibiotic may be effective in such cases.

Erythromycin is the drug of choice for patients sensitive to penicillin. A dose of 44 mg/kg up to 1 gm/day in older children and adults for 10 days is effective. Oral cephalosporins for 10 days are acceptable alternatives for penicillin-allergic patients. However, they are more expensive and should not be used in patients with immediate hypersensitivity to penicillin⁵.

Treatment of streptococcal carriers:

Streptococci may persist in the nasopharynx for a long period following untreated infection. Streptococcal carrier rate in children is high. In one study it was found that the healthy carriers do not commonly spread the organism to others. A carrier is more likely to be dangerous if large number of organisms are present and if strain is typable. It is probably wise to limit the

treatment of carriers to those who have evidence of recent infection, specially if they are members of a rheumatic family.

Prevention of streptococcal infection in non-rheumatic individuals

Treatment of family contacts:

Ideally throat cultures should be obtained and family members with significantly positive cultures be given a full course of treatment. The remainder of the family can be placed on a prophylactic regimen. If cultures are not feasible, therapy can probably be safely withheld in non-rheumatic families until symptoms and signs of illness appear.

Treatment in epidemics:

Outbreaks of virulent streptococci infections are common in military populations. They are often followed by high attack rate of RF. A number of studies have shown that mass penicillin prophylaxis of 1.2 megaunits to all members of the population affected can effectively abort epidemics and reduce the carrier rate.

Streptococcal Immunization

Active immunization would be preferable to antimicrobials in diseases characterised by repeated recurrences. The limitation of both primary and secondary prophylaxis might be overcome if an effective biologic method of prevention could be developed. The history of these attempts and the current status of streptococcal M protein vaccines are extensively reviewed⁸.

In a recent paper Stollerman⁹ is commenting to make a reasonable good case for pushing on with the long quest for safe and effective artificial immunization against group A streptococci for populations who may need some approach other than either wholesale improvement of living standards or complete dependence upon antibiotics. Such population might include 3/4 of the world's

population in areas where RHD is still a major public health problem; population even in affluent countries, in which sporadic RF still occurs in setting of poverty and poor housing. The many rheumatic subjects who do not know when to stop continuous dependence upon antibiotics, and the predisposed individual to RF following streptococcal infections. It would appear only a matter of time before the precise peptide fragments representing type-specific surface determinants is identified, against which immunization is carried out. In time, a polyvalent vaccine composing of small peptides effective against a variety of streptococcal M types is provided. Obviously, a polyvalent vaccine containing the prevalent rheumatogenic strains is a given population is valuable⁹.

Secondary Prevention

Defined as the prevention of RF recurrences in individuals with well documented history of RF or rheumatic chorea or those who have definite RHD.

Case Finding :

The need for case finding of RHD has led to introduction of various methods of screening population such as single lead (V3R) ECG, a 70 mm chest XR and limited physical examination which has to be of greatest sensitivity and specificity. Screening of school children for heart disease should be done whenever possible.

Diagnostic Facilites :

The diagnosis must depend on revised Jones criteria (Table I) which must be applied properly in clinical practise.

Table I Jones Criteria (revised)¹⁰

Major manifestations	Minor manifestations
Carditis	Fever
Polyarthrititis	Arthralgia
Chorea	Previous rheumatic fever or rheumatic heart disease
Erythema marginatum	Elevated ESR or positive CRP, Prolonged P.R interval
Subcutaneous nodules	

plus supporting evidence of preceding streptococcal infection: history of recent scarlet fever, positive throat culture for group A streptococcus, etc.

Erroneous diagnosis can be avoided by periodic re-evaluation of rheumatic patients to identify those whose original diagnosis do not meet the revised Jones Criteria and who should therefore be removed from prophylaxis^{4,8}.

This procedure has been referred to as "delabelling" of rheumatic patients. Overdiagnosis can also be reduced by encouraging physicians to diagnose patients as "rheumatic suspects" when the diagnosis remains questionable even after properly applying the Jones criteria. Such tentative diagnosis warrants prompt institution of the prophylaxis, but also provides for thorough re-evaluation of the diagnosis after specific interval⁸.

Prophylactic Methods :

Once an appropriate drug has been chosen it must be administered regularly as prescribed. Comparative studies indicate that continuous prophylaxis with 1/M penicillin is associated with lower rate of RF recurrences than is prophylaxis with oral preparation^{11,12}. In the Irvington House Studies, a recurrence rate of almost 1 per 25 patient years (10 times that of 1/M benzathine penicillin) was observed in patients with oral medication⁴. This superiority may result entirely from the certainty of administration assured by monthly injections of penicillin. Some clinics use 1/M penicillin prophylaxis exclusively in their secondary prevention programmes. Some recommended that benzathine Penicillin is the most effective form of continuous prophylaxis⁴. An attack rate of less than one recurrence per 250 patient-years was documented in patients using this form of prophylaxis in the extensive studies reported by the Irvington House group¹³.

Duration of prophylaxis:

American Heart Association (A.H.A.) recommends life long prophylaxis and exception must be made on

individual basis. Others commented that it has been difficult to establish a general recommendation concerning the duration of prophylaxis because of the numbers of variables that influence the attack rate of recurrences following streptococcal infections. Although risk of recurrences decline with age and with increased interval from the last attack, a relatively high recurrence rate per infection persists for a very long time, 5-10 years or more. Exception to instituting or maintaining prophylaxis should be in adults only and then only after assessing the risk of high exposure to streptococcal infection.

Patients with significant degrees of RHD, with a history of repeated recurrence or having had a recent attack, require consideration before discontinuing prophylaxis, a decision that must be regarded as calculated risk⁴. Majeed et al¹³ reported that if the host escapes carditis in the initial attack, he/she will continue to do so in subsequent recurrence; however, if the heart is involved in the initial attack, recurrence can inflict cardiac damage. In the view of their findings they believed that regimen should be selective; those patients with cardiac involvement should continue to be treated with probably an indefinite prophylaxis, while those patients who escaped carditis in the initial attack should maintain prophylaxis for five years. The duration of five years as they said is admittedly arbitrary but it is based on earlier observation that recurrences are most common during the first few years after the index attack. One attack of chorea is an indication for long term prophylaxis, perhaps for life¹⁴.

Case Registries :

A case registry represents an inventory of patients with certain type of chronic problem to facilitate their care and supervision. The basic objective of a registry for RF is to identify promptly those who fail to maintain their prophylaxis and medical supervision, so that they may be restored quickly to the programme through effective follow up system. Although most registries cannot serve as sources reliable incidence and prevalence data, they may provide a gross estimate of the magnitude

of RF problem in the community. In addition, it increases public and professional awareness of the significance of RF problem, and focuses interest and attention of methods of control. Experience has demonstrated that a registry must include a mechanism for the verification of diagnosis if its data are to be of value. The verification procedure itself has proved to be a valuable measure for physician education in many communities. Although the prime objective of any registry is to maintain RF patients on programme of continuous prophylaxis, in each instance the mechanics of the registry must be individualized to fit the needs of the community. A mechanism of secondary prevention programme evaluation should be included and its results continuously applied to improvement of the programme and its services.

Failures in Secondary Prevention :

Failures in secondary prevention are mostly due to dropouts from the established prophylaxis programme. It is difficult to convince a person who has recovered from his acute attack, and to convince his family as well, that although he is free of symptoms, he remains in a high risk group in regard to recurrent attacks and future cardiac damage. In order to solve his problem, more time and personal attention must be given to each patient by the physician and other clinic personnel. If the physician's attitude towards the mother indicated confidence in her reliability, strict adherence to prophylaxis was more likely. Awareness of the importance of his attitude and manner may enable the physician to modify his behaviour so as to encourage greater patient co-operation.

Adolescent rheumatic heart patient is special because the patient continues to pose problems to his prophylaxis programme. This type of patient should be tackled in a special way taking in mind the problems of adolescence. Only by awareness and understanding the adolescent and his problems and by constructing special way to provide his health services, will greater success be achieved in maintaining this high risk group in continuous prophylaxis⁸.

Community Education :

For secondary prevention, public education should be directed primarily to families having a member who has had RF. The community as a whole should be informed, however, of the extent of this problem since, from the practical standpoint broad community support is essential for its active maintenance. Such a programme will have to compete in a priority battle for support with other health programmes in the community. Thus, a strong base of community interest is vital¹.

Tertiary Prevention :

It is the prevention of further disability and death through application of medical and surgical procedures. The so called tertiary prevention includes measures of adequate treatment of patients during acute attacks, correction of heart failure, valvotomy and valve replacement etc¹⁵. Such facilities, however, are not yet available to more than a minority of patients in most countries. They represent a necessary, but most expensive approach that attempts to compensate, in part for the failure to prevent the disease in the first place.

The Role of Public Health Nurses :

Every phase of RF prevention requires the participation of a public health nurse for maximal effectiveness. The entry into homes is often an asset unique to a public health nurse, enables her to occupy an indispensable position in successful disease prevention and central programme. Primary prevention, for example; requires early case finding of streptococcal infection and this is most effectively performed by district public health nurses, who serve a segment of the population which usually does not receive adequate medical care. During hospitalization, she may assist the family by allaying their fears, assist in financial and personal problems, she helps in easing the transition to a convalescent home. She can act as health educator, supplying necessary knowledge and motivation for a patient to continue prophylaxis. In some instances the nurse may arrange transportation to the clinic for patients who would otherwise not attend. She may observe features in the home situation detrimental to the patient's health

and remedy them. She may help in securing services such as home teaching, when indicated. When residual physical disability exists, she reassures the patient and his family explains plans of medical care of long range outlook and assists in necessary readjustment.

The Role of Social Workers :

The social worker has an essential role in providing comprehensive health service to rheumatic patients. The social worker plays three main roles, the major of which is handling the non-compliant patient who could not be persuaded by the physician or the nurse. The second role is to provide assistance and advise in paramedical and personal questions related to illness, and lastly to provide advises to families for problems not related to illness such as housing and employment. The social worker's job is concerned with the attitude of the patient and his family towards illness and medical personnel, his relationship with other members of the family, the patient's personality, interest, anxieties, his school achievement, and his school-relationship with peers. The social worker collects information about the patient's living conditions and income, as this may be of help in the long term caring of the patient. The social worker evaluates the family stability and this may help to decide where convalescence should take place, whether at home or at a convalescent unit.

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