Scarlet Fever (Scarlatina) and Rheumatic Fever in Children — Symptoms and Treatment

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Scarlet fever and rheumatic fever are two possible complications of streptococcus pyogenes induced pharyngitis. Scarlet fever usually presents with a characteristic rash during the pharyngitis illness itself. Rheumatic fever presents with arthritis, carditis, skin rash, and fever a couple of weeks after a bout of streptococcal pharyngitis. Throat swab cultures are useful in the confirmation of the diagnosis of scarlet fever. Erythrocyte sedimentation rate and c-reactive proteins are usually elevated in rheumatic fever. Antibiotic treatment with penicillin, or first generation cephalosporin is indicated in both conditions. Aspirin administration is essential in rheumatic fever.
Definition of Scarlet Fever and Rheumatic Fever

Scarlet fever and rheumatic fever are two conditions associated with bacterial pharyngitis especially streptococcus pyogenes pharyngitis.

Scarlet fever is a bacterial disease that is caused by group A streptococcal bacteria. It is clinically defined as the presence of streptococcal pharyngitis with a typical skin rash. It is usually a mild disease but requires treatment for prophylaxis of more severe diseases.

Rheumatic fever is a connective tissue inflammatory disease that occurs post Group A streptococcal pharyngitis. It arises as a complication of streptococcal pharyngitis or streptococcal skin infections and affects the heart, joints and central nervous system.

Scarlet fever

- Red confluent “sandpaper” rash
- Hyperemic lines in axillae and groin (“Pastia’s lines”)
- Strawberry tongue
- Peeling fingers and toes

Epidemiology of Scarlet Fever and Rheumatic Fever in Children

Both of these conditions are found to be more common in children than in adults. Streptococcal pharyngitis is responsible for at least 20% of cases of pharyngitis in children. Children aged between 5 and 15 years constitute the majority of the cases of streptococcus pyogenes pharyngitis.

Streptococcal pharyngitis which is associated with rheumatic and scarlet fever is found to be more common in winter and spring. School going children are mostly at risk, hence emphasizing the role of schools for the transmission of the pathogen.
The most likely transmission mechanism for streptococcus pyogenes is airborne. Patients with pharyngitis who are coughing or sneezing can have the bacteria in their droplets. Exposure to infected individuals is therefore responsible for most cases of streptococcal pharyngitis.

Rheumatic fever incidences in the developed countries are very rare since the introduction of penicillin in the last few decades. In developing countries, rheumatic fever is the most common cause of heart disease in children. The diseases are more common in the developing world with poor response to infective disease and their management.

While the incidence of rheumatic fever nowadays in the developed world is believed to be very low, we do not have any conclusive epidemiological studies. In the developed world, the incidence of rheumatic fever is 0.5 per 100,000 children compared to 282 per 100,000 in the developing world.

Pathophysiology of Scarlet Fever and Rheumatic Fever in Children

It is easier to study the pathophysiology of these two conditions separately. Streptococcus pyogenes strains that produce the M substance are more likely to be associated with symptomatic pharyngitis and acute tonsillitis. These same strains are also more likely to cause scarlet fever.

**Scarlet fever** is characterized by a widespread rash that is believed to be related to an immune response to superantigens produced by streptococcus pyogenes including substance M. Other possible pathologic factors include pyrogenic exotoxins, which are produced by streptococcus pyogenes.

**Rheumatic fever** is at times preceded by an upper respiratory tract infection that is caused by streptococcus pyogenes. One important difference between scarlet fever and rheumatic fever is the timing of onset of symptoms. While the rash in scarlet fever is usually present during the stage of acute pharyngitis, patients with rheumatic
fever might present with symptoms suggestive of heart, joint or central nervous system involvement few weeks after the onset of pharyngitis.

The exact pathogenesis mechanism of rheumatic fever is still poorly understood. Despite this, our current understanding of the disease makes it possible to prevent the condition. The most likely pathology in rheumatic fever is the activation of an inflammatory immune response to certain antigens produced by streptococcus pyogenes that resemble our own cellular surface antigens.

Therefore, inflammation of the myocardium, heart valves, joints, and the central nervous system is thought to be immune mediated. Because of this understanding, we currently can prevent the development of rheumatic fever by early and prompt antibiotic therapy of streptococcus pharyngitis.

Clinical Presentation of Scarlet Fever and Rheumatic Fever in Children

Children with scarlet fever usually complain of acute pharyngitis that is associated with a typical rash. The rash starts within the onset of pharyngitis and is usually on the trunk but can spread to the extremities. Patients with scarlet fever have flushed faces. The rash consists of many small papules on the skin that resemble sand.

On examination, a red tongue can be seen in patients with scarlet fever. This sign is known as strawberry tongue. The rash in scarlet fever is known to desquamate later within one week or so.

As both conditions are known to be associated with streptococcus pharyngitis, differentiation of viral versus bacterial pharyngitis based on clinical scoring was studied. The presence of high grade fever, absence of cough, presence of tonsillar exudates and painful cervical lymphadenitis is suggestive of streptococcal pharyngitis.

Patients with rheumatic fever are usually diagnosed based on a classification system that takes into account major and minor manifestations. Major manifestations of rheumatic fever include carditis, polyarthritis, chorea, which is a characteristic movement disorder, the presence of subcutaneous rheumatic nodules and a characteristic rash known as erythema marginatum.

Minor criteria for the diagnosis of rheumatic fever include fever, joint pain without evidence of arthritis, elevated erythrocyte sedimentation rate or c-reactive protein and a
prolonged PR interval on electrocardiogram.

The diagnosis of rheumatic fever is based on the documentation of the presence of a recent streptococcal pharyngitis illness plus the presence of two of the major criteria or one of the major criteria and two of the minor ones.

Due to cardiac involvement in rheumatic fever, patients can present with tiredness, dyspnea and chest pain. Patients with rheumatic fever usually have migrating arthritis where it seems that once a joint improves another becomes inflamed.

**Diagnostic Workup for Scarlet Fever and Rheumatic Fever in Children**

Patients presenting with symptoms and signs suggestive of scarlet fever should undergo a **positive culture of a throat swab** looking for streptococcus pyogenes. **Rapid antigen detection tests** are also available in the evaluation of patients with scarlet fever but their accuracy remains below that of throat culture.

![Streptococcus pyogenes.](https://via.placeholder.com/150)

Patients who present with symptoms suggestive of rheumatic fever but who did not have any documentation of a recent streptococcal pharyngitis in the last two weeks might benefit from **serologic testing**. The detection of **specific antibodies against the SLO or DNase B antigens**, which are related to streptococcus pyogenes can be considered as evidence of a recent streptococcal pharyngitis and help with the diagnosis of rheumatic fever.

Patients with rheumatic fever should also undergo **erythrocyte sedimentation rate** and **c-reactive protein** measurement. These two acute phase reactants are usually elevated in rheumatic fever. **Electrocardiography** is also indicated to look for possible specific signs of rheumatic fever such as a prolongation of the PR interval. **Ventricular arrhythmias** may be seen on electrocardiography as a consequence to myocarditis.

Patients with suspected rheumatic fever should undergo **echocardiography** assessment of the heart. If the echocardiogram does not reveal any heart problems such as impaired contractility, or valvular abnormalities, the test should be repeated again after two weeks. If the echocardiography is still normal at two weeks, no further cardiac follow-up is indicated.

Patients who have an **abnormal echocardiogram** should be **monitored closely** as the
most common cause of death in rheumatic fever is cardiac related. The test should be repeated at six weeks to look for any signs of functional improvement of the heart. If the patient’s condition becomes worse, urgent repeat echocardiography is advised.

Follow-up is recommended at three months and twelve months after the diagnosis of cardiac abnormalities related to rheumatic fever. The most commonly identified cardiac abnormalities are aortic or mitral valve regurgitation.

Diagnosis of rheumatic fever

- Evidence of recent strep infection (rapid strep, ASO titer, or anti-DNAse B)
- Two major criteria, one minor criterion
- One major criterion, two minor criteria
- Sydenham’s chorea alone

Treatment of Scarlet Fever and Rheumatic Fever in Children

The antibiotic therapy of scarlet fever is important as it decreases the duration of the illness, prevents rheumatic fever, and prevents other pharyngitis related complications.

Amoxicillin or penicillin are the antibiotics of choice in the management of scarlet fever. One of these two antibiotics should be used for ten days for optimum outcome. Patients with scarlet fever who are allergic to penicillins should receive cephalexin or cefadroxil for ten days.

Amoxicillin or penicillin

- Treat immediately for 10 days to prevent any ongoing antibody production from lingering infection.
- Patients with heart disease may require prophylaxis with daily amoxicillin or monthly penicillin IM for decades.
- Use supportive treatment for heart disease and/or transplant.

One advantage of azithromycin over amoxicillin and cephalosporins is the shorter duration of treatment. Azithromycin can be used once a day for five days with similar results. The amount of evidence, however, supporting the use of azithromycin for streptococcal pharyngitis is inferior compared to what is available for penicillins or cephalosporins.

Patients with rheumatic fever usually do not have an ongoing streptococcal infection, therefore the role of antibiotics is less clear. Regardless, a single dose of intramuscular benzathine penicillin or erythromycin are recommended in all patients presenting
with rheumatic fever. The aim of this approach is to eradicate streptococcus pyogenes from the throat.

Patients with rheumatic fever usually complain of arthritis and joint pain. Salicylic acid, i.e. aspirin, administration is indicated in the management of inflammation induced by rheumatic fever. More recently, it was shown that naproxen is a safer option and as effective as aspirin for the management of rheumatic fever.

Patients with severe myocarditis might benefit from corticosteroids administration. Bed rest is usually indicated in patients with rheumatic fever because joint pain is usually very severe. Patients with arthritis alone without any evidence of cardiac involvement should have bed rest for one to two weeks. On the other hand, patients with mild to moderate carditis should have bed rest for one to two months.

Patients with severe myocarditis and myocardial edema might benefit from prolonged bed rest for two to four months. This group of patients might develop congestive heart failure.

References

Acute rheumatic fever in children: recognition and treatment via nih.gov
Pharyngitis and Scarlet Fever via nih.gov

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