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

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 streptococcal pharyngitis. Throat swab cultures are useful in confirming 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 for both conditions. Aspirin administration is essential for 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 an associated skin rash. Scarlet fever is usually a mild disease but requires treatment for prophylaxis of more severe diseases. Rheumatic fever is a connective tissue inflammatory disease that occurs after 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 more common in children than in adults. Streptococcal pharyngitis is responsible for at least 20% of cases of pharyngitis in children. Children aged 5-15 years constitute the majority of *Streptococcus pyogenes* pharyngitis cases.

*Streptococcal pharyngitis* is more common in winter and spring. School-going children are most at risk, as schools are often where the pathogen is transmitted.

The most likely transmission mechanism for *Streptococcus pyogenes* is airborne. Patients with pharyngitis who are coughing or sneezing can spread the bacteria through their droplets. Exposure to infected individuals is therefore responsible for most cases of streptococcal pharyngitis.
Rheumatic fever incidences in 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. Such diseases are more common in the developing world because of poor response to and management of infectious diseases.

While the incidence of rheumatic fever in the developed world is believed to be very low, there are no 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 sometimes 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 the 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 a few weeks after the onset of pharyngitis.

The exact pathogenesis of rheumatic fever is still poorly understood; however, 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, the development of rheumatic fever can be prevented by early and prompt antibiotic therapy of streptococcal 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 rash. The rash starts with the onset of pharyngitis and is usually on the trunk but can spread to the extremities. Patients with scarlet fever also have flushed faces. The rash consists of many small papules on the skin that resemble sand. The rash will usually desquamate within about one week. Upon examination, the patient presents with a red tongue, known as strawberry tongue.

As both conditions are associated with streptococcal pharyngitis, differentiation of viral versus bacterial pharyngitis based on clinical scoring has been studied. The presence of a high-grade fever, absence of a cough, presence of tonsillar exudates, and painful cervical lymphadenitis suggest streptococcal pharyngitis, rather than viral pharyngitis.

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

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 the electrocardiogram.

The diagnosis of rheumatic fever is based on the documentation 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.
Diagnostic Workup for Scarlet Fever and Rheumatic Fever in Children

Patients presenting with symptoms that suggest scarlet fever should undergo a throat swab culture to evaluate for *Streptococcus pyogenes*. Rapid antigen detection tests are also available to evaluate patients with scarlet fever, but they are less accurate than throat cultures.

![Image: Streptococcus pyogenes. By CDC. License: Public Domain](image)

Patients who present with symptoms that suggest rheumatic fever but who have not had streptococcal pharyngitis in the last two weeks may 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 evidence of recent streptococcal pharyngitis and help diagnose rheumatic fever.

Patients with rheumatic fever should also undergo an erythrocyte sedimentation rate and c-reactive protein measurement. These two acute phase reactants are usually elevated in patients with rheumatic fever.

Electrocardiography is also indicated to look for possible signs of rheumatic fever, such as a prolonged PR interval. Ventricular arrhythmias may be seen on electrocardiography as a consequence of myocarditis. 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 causes of death in rheumatic fever patients are heart-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 common cardiac abnormalities are aortic or mitral valve regurgitation.

**Diagnosis of rheumatic fever**

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

Treatment of Scarlet Fever and Rheumatic Fever in Children

The **antibiotic therapy** for scarlet fever is important, as it decreases the duration of the illness, prevents rheumatic fever, and prevents other pharyngitis-related complications. Amoxicillin and penicillin are the antibiotics of choice in the management of scarlet fever. One of these antibiotics should be used for 10 days for optimal outcome. Patients with scarlet fever who are allergic to penicillins should receive cephalexin or cefadroxil for 10 days.

**Amoxicillin or penicillin**

- Treat immediately for 10 days to prevent ongoing antibody production from any lingering infection.
- Patients with heart disease may require prophylaxis with daily amoxicillin or monthly penicillin intramuscularly 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. There is less evidence, however, to support the use of azithromycin for streptococcal pharyngitis compared with 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 is 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, the administration is indicated to manage inflammation induced by rheumatic fever. More recently, research has shown that naproxen is a safer option and as effective as aspirin for the management of rheumatic fever symptoms.

Patients with severe myocarditis may benefit from corticosteroid administration. Bed rest is usually indicated in patients with rheumatic fever because joint pain is often severe. Patients with arthritis alone and no evidence of cardiac involvement should be prescribed bed rest for 1–2 weeks. On the other hand, patients with mild to moderate carditis should be prescribed bed rest for 1–2 months. Patients with severe myocarditis and myocardial edema may benefit from prolonged bed rest for 2–4 months. These patients may develop congestive heart failure.

**References**


[Pharyngitis and Scarlet Fever](https://nih.gov) via nih.gov