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Rheumatic Fever And Post-streptococcal Reactive Arthritis

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2. DIAGNOSIS AND THERAPY

2.1 How is it diagnosed?

Clinical signs and investigations have special importance, because there is no specific test or sign for the diagnosis. Clinical symptoms of arthritis, carditis, chorea, skin changes, fever, abnormal laboratory tests for streptococcal infection and changes in the conduction of the heartbeat, as seen on an electrocardiogram, can help establish the diagnosis (so called Jones criteria). Evidence of a preceding streptococcal infection is generally needed to make the diagnosis.

2.2 Which diseases are like rheumatic fever?

There is a disease called "post-streptococcal reactive arthritis" that also occurs after streptococcal pharyngitis, but which features arthritis of longer duration and which has a lower risk of carditis; antibiotic prophylaxis may be indicated. Juvenile idiopathic arthritis is another disease resembling rheumatic fever, but the duration of arthritis is longer than 6 weeks. Lyme disease, leukaemia, reactive arthritis caused by other bacteria or viruses may also present with arthritis. Innocent murmurs (common heart murmurs with no heart disease), congenital or other acquired heart diseases can be misdiagnosed as rheumatic fever.

2.3 What is the importance of tests?

Some tests are essential for the diagnosis and follow-up. Blood tests are

useful during attacks to confirm the diagnosis.
As in many other rheumatic diseases, signs of systemic inflammation are seen in nearly all patients, except those with chorea. In most patients, there is no sign of throat infection and the streptococcus bacteria in the throat are cleared by the immune system by the time of disease onset. There are blood tests to detect streptococcal antibodies, even if the parents and/or the patient are not able to recall the signs of throat infection. Rising titres (levels) of these antibodies, known as anti-streptolysin O (ASO) or DNAse B, can be detected by blood tests carried out 2-4 weeks apart. High titres indicate a recent infection, but there is no proven relationship with disease severity. However, these tests yield normal results in patients with only chorea, making this diagnosis tricky. Abnormal ASO or DNAse B test results mean prior exposure to the bacteria stimulating the immune system to produce antibodies, and by itself, does not make the diagnosis of rheumatic fever in patients without symptoms. Antibiotic treatment is therefore generally not necessary.

2.4 How to detect carditis?

A new murmur, resulting from heart valve inflammation, is the most common feature of carditis and is usually detected when a physician auscultates (listens to) the heart sounds. An electrocardiogram (an assessment of heart’s electrical activity registered on a paper strip) is useful to ensure the extent of heart involvement. Chest X-rays are also important to check for enlargement of the heart.
Doppler echocardiogram or heart ultrasound is a very sensitive test for carditis. All these procedures are absolutely painless and the only discomfort is that the child must keep still for a while during the test.

2.5 Can it be treated/cured?

Rheumatic fever is an important health problem in certain areas of the world, but it can be prevented by treating streptococcal pharyngitis as soon as it is recognized (i.e. primary prevention). Antibiotic therapy started within 9 days of onset of pharyngitis is effective in preventing acute rheumatic fever. Symptoms of rheumatic fever are treated with non-steroidal anti-inflammatory medications.
Research is currently being carried out to produce a vaccine that can

protect against streptococcus: prevention of the initial infection would provide protection against the abnormal immune reaction. This approach might prevent rheumatic fever in the future.

2.6 What are the treatments?

During the past several years, there have been no new treatment recommendations. While aspirin has remained the mainstay of therapy, the exact action of effect is unclear; it appears to be related to the anti-inflammatory properties. Other non-steroidal anti-inflammatory drugs (NSAIDs) are recommended for arthritis for 6-8 weeks or until it disappears.

For severe carditis, bed rest and, in some cases, oral corticosteroids(prednisone) are recommended for 2-3 weeks, tapering the drug off gradually after the inflammation is controlled by observing the symptoms and by blood tests.

In the event of chorea, parental support for personal care and school tasks may be required. Drug treatment for chorea movement control with steroids, haloperidol or valproic acid may be prescribed with close monitoring for side-effects. Common side effects are sleepiness and trembling that can be easily controlled by dose adjustment. In a few cases, chorea may last for several months despite adequate treatment. After the diagnosis is confirmed, long-term protection with antibiotics is recommended to avoid recurrence of acute rheumatic fever.

2.7 What are the side effects of drug therapy?

In terms of short-term symptomatic treatment, salicylates and other NSAIDs are usually well tolerated. The risk of a penicillin allergy is quite low, but its use must be monitored during the first injections. The main considerations are the painful injections and possible refusal by patients who fear pain; therefore, education about the disease, topical anaesthetics and relaxation before injections are recommended.

2.8 How long should secondary prevention last?

The risk of relapse is higher during the 3-5 years after onset and the risk of developing carditis damage increases with new flares. During this time, regular antibiotics treatment for preventing new streptococcal

infections is recommended for all patients who have had rheumatic fever, regardless of the severity, as mild forms may flare as well. Most physicians agree that antibiotic prevention should continue for at least 5 years after the last attack or until the child is 21 years old. In the event of carditis without heart damage, secondary prophylaxis is recommended for 10 years or until the patient is 21 years old (whichever is longer). If heart damage is present, 10 years of prophylaxis are recommended, or until the age of 40 - or later in life if the disease is complicated by valve replacement.
Prevention of bacterial endocarditis with antibiotics is recommended for all patients with heart valve damage undergoing dental work or surgery. This measure is necessary because bacteria can move from other sites of the body, especially from the mouth, and cause heart valve infection.

2.9 What about unconventional/complementary therapies?

There are many complementary and alternative therapies available and this can be confusing for patients and their families. Think carefully about the risks and benefits of trying these therapies as there is little proven benefit and they can be costly in terms of time, burden to the child and money. If you want to explore complementary and alternative therapies, it is wise to discuss these options with your paediatric rheumatologist. Some therapies can interact with conventional medications. Most doctors will not be opposed to complementary therapies, provided you follow medical advice. It is very important not to stop taking your prescribed medications. When medications, such as corticosteroids, are needed to keep the disease under control, it can be very dangerous to stop taking them if the disease is still active. Please discuss medication concerns with your child’s doctor.

2.10 What kind of periodic check-ups are necessary?

Regular check-ups and periodic tests may be required during long-term disease course. Closer follow-up is recommended in cases where there is carditis and chorea. After remission of the symptoms, a supervised schedule for preventive treatment and long-term follow-up under supervision of a cardiologist looking for late heart damage is recommended.

2.11 How long will the disease last?

Acute symptoms of the disease recede over several days to weeks. However, the risk of recurrent attacks of acute rheumatic fever persists and heart involvement can cause life-long symptoms. Ongoing antibiotic treatment to prevent recurrence of streptococcal pharyngitis is necessary for many years.

2.12 What is the long-term evolution (prognosis) of the disease?

Relapses of symptoms tend to be unpredictable in time and severity. Carditis in the first attack increases damage risk, although complete healing may follow carditis in some cases. Most severe heart damage requires heart surgery for valve replacement.

2.13 Is it possible to recover completely?

Complete recovery is possible, unless carditis resulted in severe heart valve damage.