



The Importance of Early Diagnosis for the Treatment of Rheumatic Heart Disease

Thalita P. M. Alineri¹; Délio T. M. Malaquias¹; Juliana F. B. Paschoal¹; Lucimara Pigaiani¹; Marcus B. Fonseca¹; Isabelle P. Santos¹; Wellington S. P. Cunha¹; Juliana F. B. Paschoal¹; Cristiana N. O. Beloto¹; Danilo A. O. Panebianco¹; Isabelle P. Santos¹; Ana Carolina C. Cruz¹; Lucas S. Vieira¹; Priscilla E. A. Torres¹; Leticia S. Assis¹; Heloise B. O. Rodrigues¹; Gianluca F. C. Sganzella¹; Adriana F.V. Delgado¹; Larissa R. S. Martins¹; Arthur G.S.C. Monteiro¹; Larissa A. Abreu¹; Alberto E. Yamane¹; Cristiano de Melo¹; Talita R. Q. Lopes¹; Samantha R.G. Sanches¹; Caroline P. Golin¹; Elisa F. Prezotto¹; Leonardo T. Silva¹; Aghata A.M. Faria¹; Hiromi M. K. Fujishima²; Thiago G. Trigueiro²; Pedro N. S. Costa²; Thiago A.R.Bezerra¹⁻³

¹ Medical student. University of Ribeirão Preto. Guarujá, São Paulo, Brazil.

² Medical student. Potiguar University. Natal. Rio Grande do Norte, Brazil.

³ Medical student. University of Ribeirão Preto. Guarujá, São Paulo, Brazil. Bachelor in Physical Education. Federal University of São Carlos, São Paulo, Brazil. PhD in Medical Sciences. University of São Paulo. Ribeirão Preto, São Paulo, Brazil.

DOI: <https://doi.org/10.55248/gengpi.4.1123.112903>

ABSTRACT

Introduction: Rheumatic Fever (RF) is characterized by being an inflammatory disease that occurs after bacterial tonsillitis caused by Streptococcus and inadequately treated. The infection is clinically presented by fever, sore throat, enlarged lymph nodes and intense redness, red dots or plaques of pus in the throat. The disease can spread to the joints, heart and brain, leaving severe cardiac sequelae with lifelong consequences and possibly leading to death. The disease occurs in outbreaks, if not prevented, and with each outbreak the chance of serious heart damage increases. Although RF can occur at all ages, the 5 to 15 age group is the most affected. Objectives: To carry out a literature review on early diagnosis, treatment and new parameters related to rheumatic heart disease. Literature review: Prevention of the disease is considered relatively easy, and depends solely on the appropriate treatment of tonsillitis, which can be done with the administration of a single dose of benzathine penicillin. However, once RF is established, it can cause severe heart damage, leading to repeated hospitalizations, complex cardiovascular surgery and drug treatment that is difficult to manage, such as the use of anticoagulants for the rest of one's life, influencing the working capacity of patients and their guardians. Final considerations: Early diagnosis of cases of bacterial tonsillitis among school-age children and referral of these suspected cases for the most appropriate treatment is of fundamental importance.

Keywords: Rheumatic fever; Bacterial tonsillitis, Carditis.

INTRODUCTION

Rheumatic heart disease is a disease that especially affects the heart valves, resulting from repeated throat infections (tonsillitis) caused by streptococcus bacteria (ASMARE et.al, 2021).

According to Athayde (2015), around 33 million people in the world are affected by rheumatic heart disease - the number of annual deaths reaches 320 thousand.

Pharyngotonsillitis caused by rheumatic fever (RF) and chronic rheumatic heart disease (CRC) is caused by group A beta-hemolytic streptococcus (ASMARE et.al, 2021).

It is common to associate RF with poverty and poor living conditions. Thus, despite the recognized reduction in the incidence of RF in recent decades in developed countries, with a consequent reduction in the prevalence of CRC, RF remains a major public health problem, especially in developing countries (ATHAYDE, 2015).

According to Asmare et al (2021), RF can affect children and young adults in particular. The most fearsome manifestation is carditis, which is responsible for chronic, often disabling sequelae in the early stages of life, generating high social and economic costs.

It is important to create effective strategies for the prevention and treatment of streptococcal pharyngotonsillitis. The health actions developed to date have proved insufficient to adequately control RF in Brazil (CABRAL, et.al, 2019).

Studies by Cabral et.al (2019) have shown that in developed and developing countries, pharyngotonsillitis and impetigo are the infections most frequently caused by group A beta-hemolytic streptococcus (GABS). Their incidence varies from country to country and, within the same country, varies according to the different regions, basically depending on the age of the individual, socioeconomic conditions, environmental factors and the quality of health services.

According to Cabral et.al (2019), it is very difficult to determine the incidence of bacterial pharyngotonsillitis caused by EBGA. This would require trained professionals and tests to detect the presence of the bacteria, as well as an efficient information system.

RF has a universal distribution, but with marked differences in incidence and prevalence rates between different countries, and is the main cause of acquired heart disease in children and young adults in developing countries (CARAPETIS, et.al, 2013).

According to Carapetis et.al (2013), the available data basically refers to hospital admissions and interventions, and does not correspond to all diagnosed cases.

OBJECTIVES

To carry out a literature review on early diagnosis, treatment and new parameters related to rheumatic heart disease.

MATERIAL AND METHODS

This article is a literature review that seeks to identify early diagnosis, treatment and new parameters related to rheumatic heart disease, by analyzing previous studies.

A search strategy was developed based on the evaluation of an objective on the subject in question, which forms the basis of the study. This was: To identify rheumatic heart disease and its implications.

The search descriptors were selected from the Descriptors in Health Sciences (DeCS) website and then combined with the Boolean operator "AND". The databases used for the search were: PubMed and the Virtual Health Library (VHL), where cross-sectional, cohort and case-control studies were evaluated, covering the languages Portuguese and English.

In all, the result of the search in the databases using the descriptors, but without the application of filters, resulted in 112 articles available.

The divergence pointed out in relation to the type of literature filter, which could lead to a selection bias, was resolved by selecting only cross-sectional articles in the inclusion criteria. After evaluating each article individually, analyzing the title and abstract of the study and applying the inclusion and exclusion criteria, 59 articles were pre-selected.

After pre-selecting the articles, a research protocol was created which clearly illustrated the aim of the study, the data collection process and the criteria involved in including the articles.

After the analysis, 30 studies were excluded. Therefore, 29 articles were selected for this review.

LITERATURE REVIEW

A brief history

The Jones criteria, so traditional in the diagnosis of rheumatic fever, emerged in 1944 with a clear bias: they had high specificity and low sensitivity (SAXENA et.al, 2017).

The development of RF is associated with EBGA infection of the oropharynx, especially in children and adolescents. Genetic disease susceptibility factors are directly related to the development of RF and its sequelae (ROBERTS, 2015; SAXENA et.al, 2017).

Several systems with genetic polymorphism have been and are being studied in order to elucidate the onset of RF and perhaps, in the future, help prevent it in families with cases of the disease (SCHEEL, 2019).

The immune response in rheumatic disease

Carapetis et.al (2013) describes that the pathophysiological mechanisms of rheumatic fever consist of the body's autoimmune inflammatory response to group A streptococci. This reaction is due to molecular mimicry, which consists of the chemical and structural similarity between some pathogenic components and the affected tissue.

The existence of an autoimmune process in RF was postulated after the observation that lesions in the heart were associated with antibodies that recognized cardiac tissue by molecular mimicry, whose data were experimentally confirmed by Kaplan (REMENYI, et.al, 2013).

Diagnosis

The diagnosis of rheumatic fever is clinical and there are no pathognomonic signs or specific tests. Laboratory tests, although non-specific, support the diagnosis of the inflammatory process and streptococcal infection (MARIJON, et.al, 2007).

According to Martins (2022), the Jones criteria, established in 1944, were last modified in 1992 and are still considered the "gold standard" for diagnosing the first outbreak of RF. The division of the criteria into major and minor is based on specificity and not on the frequency of the manifestation. Other signs and symptoms, such as epistaxis, abdominal pain, anorexia, fatigue, weight loss and pallor may be present, but are not included among the minor manifestations of the Jones criteria.

According to Muller et al (2015), the likelihood of RF is high when there is evidence of previous streptococcal infection, determined by elevated anti-streptolysin O (ASLO) titers, as well as the presence of at least two major criteria or one major and two minor criteria. With successive modifications, the criteria have improved in specificity and lost in sensitivity due to the requirement to prove streptococcal infection.

The Jones criteria modified by the American Heart Association (AHA) in 1992 should be used to diagnose the first outbreak of the disease, while the Jones criteria revised by the WHO and published in 2004 are also used to diagnose recurrences of RF in patients with established CRC (VASCONCELOS, 2020).

Diagnosis of pharyngotonsillitis

Pharyngotonsillitis is an acute infection of the pharynx, tonsils or both. Symptoms may include a sore throat, pain in the throat, cervical lymphadenopathy and fever. Diagnosis is clinical, complemented by culture or a rapid antigen test (MARIJON, et.al, 2007).

According to Mirabel et.al (2015) 25% of these EBGA infections are subclinical manifestations. This can result in patients not seeking medical attention, as well as doctors underdiagnosing the disease.

The diagnosis of streptococcal pharyngotonsillitis allows for appropriate antimicrobial treatment and, consequently, primary prevention of RF. The differential diagnosis of streptococcal infection of the oropharynx, which accounts for approximately 30% of cases of pharyngotonsillitis, should be made, particularly with viral infections. Clinical manifestations can help in this differentiation. The diagnosis of streptococcal pharyngotonsillitis can be suggested by the presence of the clinical criteria validated by the WHO, which include: general malaise, vomiting, high fever, hyperemia and edema of the oropharynx, as well as petechiae and purulent exudate, in addition to palpable and painful cervical lymph nodes. On the other hand, the presence of a runny nose, cough, hoarseness and conjunctivitis suggest a viral infection (SZCZYGIELSKA et.al, 2018).

According to Muller et.al (2015), laboratory proof of EBGA infection is recommended. Oropharyngeal culture is the "gold standard" for diagnosing streptococcal pharyngotonsillitis and has a sensitivity of 90%-95% (WEBB, 2007).

According to Mirabel et.al (2015), when compared to culture, the rapid antigen detection test has the advantage of rapid results and also has a sensitivity of 80% and specificity of 95%.

If the clinical picture is suggestive of streptococcal pharyngotonsillitis and the rapid test is negative, an oropharyngeal culture is recommended (ROBERTS, 2015; SAXENA et.al, 2017).

Serological tests reflect a previous infection and are of no value in diagnosing acute streptococcal pharyngotonsillitis. The most commonly used tests are anti-streptolysin O (ASLO) and anti-deoxyribonuclease B (anti-DNase). ASLO titers only confirm the presence of previous streptococcal infection (SZCZYGIELSKA et.al, 2018).

Pharyngotonsillitis (FIGURE 1) are infections that affect the tonsils, posterior pharynx, soft palate and lymphoid organs in the region. They are very common infections, especially in the pediatric age group, which is why many children are absent from school and their learning is impaired (ROBERTS, 2015; SAXENA et.al, 2017).

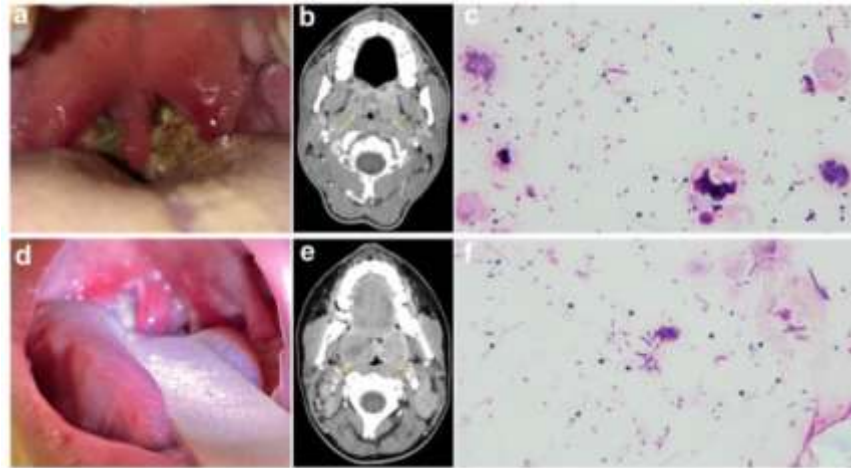


Figure 1. Necrotic pseudomembranous pharyngotonsillitis due to infectious mononucleosis and Plaut-Vincent tonsillitis in a young man (a) and a woman (d). Both had large tonsillitis without abscess on computed tomography images (b and e). The fusospirillar association visualized after Gram staining of a tonsil smear (c and f) revealed the presence of fusiform Gram-negative bacilli evocative of *Fusobacterium nucleatum* (→) and spiral Gram-negative bacilli evocative of *Treponema vincentii*. **Source:** clinicalmicrobiologyandinfection.com

According to Feitosa et al (1991), the rise in titers starts around the 7th day after infection and peaks between the 4th and 6th week, remaining high for months, sometimes even a year after infection. It is recommended that two ASLO tests be carried out 15 days apart (IB).

It has been observed that approximately 20% of patients with RF do not have elevated ASLO. It is recommended to use the standard limits of each laboratory to analyze the results (VASCONCELOS, 2020).

Major Criteria

Arthritis

Arthritis is the most common manifestation of RF, present in 75% of cases, with a self-limiting course and no sequelae. It is often the only major criterion present, especially in adolescents and adults. In cases of association with carditis, an inverse correlation has been described between the severity of the two manifestations (SZCZYGIELSKA et.al, 2018).

The differentiation between arthritis (major criterion) and arthralgia (minor criterion) is made on clinical grounds. Arthralgia means only joint pain, while arthritis is defined as the presence of edema in the joint or, failing that, by the association of pain with limitation of movement (SOUZA, 2006).

Typical RF arthritis develops asymmetrically and migratory, i.e. when symptoms improve in one joint, they appear in another. The joint condition preferentially affects the large joints, particularly those of the lower limbs. The duration of the inflammatory process in each joint rarely exceeds a week and the total condition ceases in less than a month. Arthritis is usually very painful, although it does not show intense signs of inflammation on physical examination. The response to non-hormonal anti-inflammatory drugs is rapid and the pain often disappears within 24 hours, while the other inflammatory signs cease within two to three days (WATKINS, et.al, 2018).

Although the typical pattern is observed in around 80% of cases, there are atypical presentations that require other considerations in the differential diagnosis, which include additive arthritis (progressive and simultaneous involvement of several joints, without inflammation ceasing in the previous ones), monoarthritis and involvement of small joints and the spine (ROBERTS, 2015).

The early use of non-hormonal anti-inflammatory drugs may be responsible for the absence of the migratory characteristic of arthritis, since the excellent response prevents the progression of signs and symptoms, which may result in a monoarticular condition. In these cases, discontinuing the anti-inflammatory drugs may cause the arthritis to return and allow a better assessment of the diagnosis. Small joints, such as the interphalangeal and metacarpophalangeal joints, may be involved, but there should always be concomitant involvement of large joints. Arthritis of the spine, especially the cervical spine, can be revealed by the presence of pain and limited movement (ZÜHLKE, et.al, 2013).

Post-streptococcal reactive arthritis

Post-streptococcal reactive arthritis (PSRA) affects all age groups, especially adults, with involvement of more than one joint, but does not meet the Jones criteria for diagnosing RF. The time interval between oropharyngeal infection and the onset of joint pain is around 10 days, which is shorter than in rheumatoid arthritis. Some authors consider ARPE to be part of the spectrum of RF, while others consider it to be a separate entity (ZAHARI, et.al, 2022).

ARPE is usually cumulative and persistent, involving large and/or small joints and does not respond satisfactorily to the use of salicylates or other non-hormonal anti-inflammatory agents (ZAHARI, et.al, 2022).

The degree of concomitant cardiac involvement is uncertain and probably uncommon. Given the difficulties in differentiating ARPE from rheumatoid arthritis, patients with ARPE should be considered to have RF when they meet the Jones criteria (WHITE, et.al, 2010).

Carditis

According to Mirabel et al (2015), carditis is the most serious manifestation of RF, as it is the only one that can leave sequelae and lead to death. The manifestation occurs in 40%-70% of the first outbreaks, although more recent series, in which echocardiography was used for evaluation, show higher prevalence rates.

Carditis tends to appear at an early stage and is most often diagnosed in the first three weeks of the acute phase. Cardiac involvement is characterized by pancarditis, but it is the valve lesions that are responsible for the clinical picture and prognosis (MIRABEL, et.al, 2015).

According to Feitosa et.al (1991), pericardial involvement is not common, does not occur in isolation and does not result in constriction. Pericarditis is always associated with valve damage and is diagnosed by the presence of friction and/or pericardial effusion, muffled sounds, precordial pain or discomfort. In mild cases, pericardial involvement is an exclusive finding of the echocardiographic study. Large pericardial effusions and cardiac tamponade are rare.

Myocarditis has been diagnosed on the basis of a muffled first sound, protodiastolic gallop, cardiomegaly and congestive heart failure. Despite histological and immunological evidence of myocardial involvement, heart failure is caused by valvular lesions and not by myocardial involvement. Left ventricular systolic function indices are generally preserved in the initial outbreaks (FEITOSA, et.al, 1991).

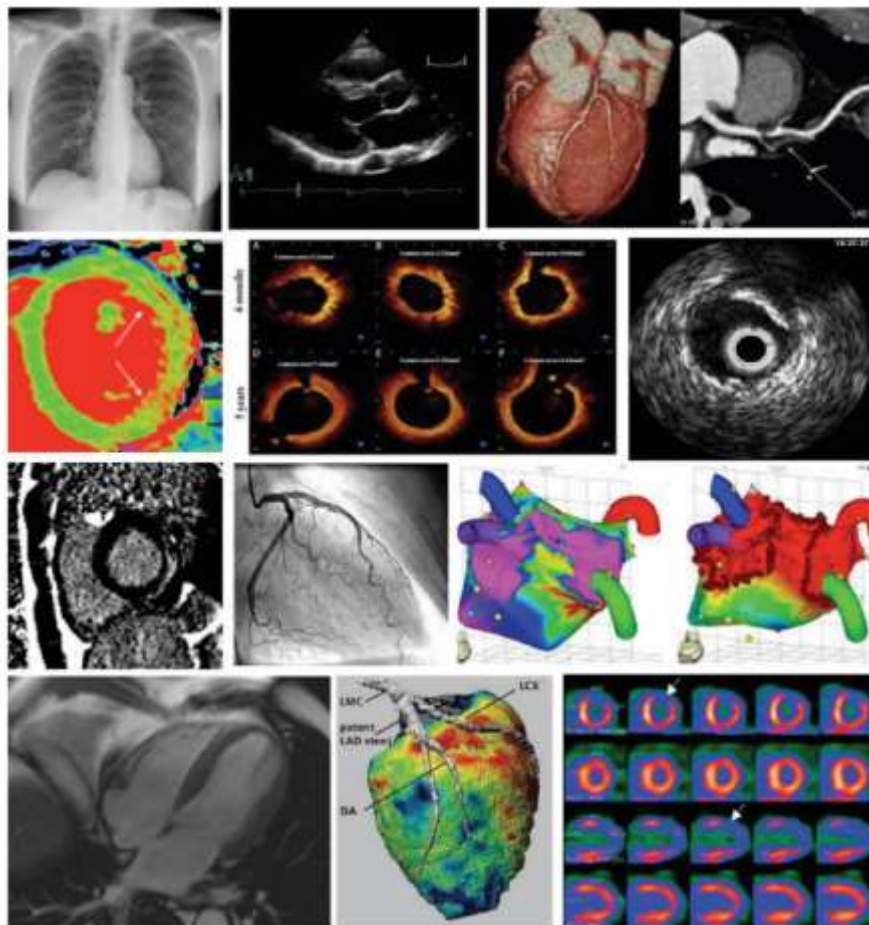


Figure 2: Cardiovascular magnetic resonance imaging in rheumatic heart disease. **Source:** [semanticscholar.org/paper/Cardiovascular-magnetic-resonance-imaging-in-heart](https://www.semanticscholar.org/paper/Cardiovascular-magnetic-resonance-imaging-in-heart)

Endocardial involvement (endocarditis/valvitis) is the diagnostic hallmark of carditis, most frequently involving the mitral and aortic valves. In the acute phase, the most frequent lesion is mitral regurgitation, followed by aortic regurgitation. On the other hand, valve stenosis occurs later in the chronic phase. It is worth noting that mitral valve regurgitation is more prone to total or partial regression than aortic regurgitation (WEBB, 2007).

According to Feitosa et al (1991), three murmurs are characteristic of the first episode and may not represent definitive valve dysfunction: mitral regurgitation systolic murmur, Carey Coombs diastolic murmur and aortic regurgitation diastolic murmur. The absence of a murmur does not rule out the possibility of cardiac involvement. Mild carditis unaccompanied by other symptoms of the disease may go unnoticed, and valve damage may only be revealed during routine medical examinations or during subsequent outbreaks.

For Gomes (2022), subclinical carditis is found in patients with isolated arthritis and/or pure chorea, without auscultatory findings of valve damage, but with a pathological pattern of regurgitation on doppler echocardiography. It is important to use strict criteria to differentiate pathological from physiological regurgitation.

According to Joseph et.al (2013), recurrent carditis is suspected by detecting a new murmur or an increase in the intensity of previously existing murmurs, pericardial friction or effusion, an increase in cardiac area or heart failure associated with evidence of previous streptococcal infection.

According to Ledos (2015), in indolent carditis, the clinical manifestations are discreet and the clinical picture has a prolonged evolution.

The studies by Marijon et al (2007) describe that the severity of carditis, depending on the degree of cardiac involvement, ranges from subclinical to fulminant. According to the clinical, radiological, electrocardiographic and dopplerecardiographic findings, carditis can be classified into the following categories.

Sydenham's chorea

According to Mirabel et.al (2015), Sydenham's chorea (SC) occurs predominantly in female children and adolescents, and is rare after the age of 20. Its prevalence ranges from 5%-36% in different reports, with an insidious onset usually characterized by emotional lability and muscle weakness that make diagnosis difficult.

Sydenham's chorea (SC) occurs predominantly in female children and adolescents and is rare after the age of 20. Its prevalence ranges from 5%-36% in different reports, with insidious onset usually characterized by emotional lability and muscle weakness that make diagnosis difficult (SZCZYGIELSKA et.al, 2018).

Subcutaneous nodules

According to Martins (2022), subcutaneous nodules are rare, present in only 2%-5% of patients, and are strongly associated with the presence of severe carditis. They are multiple, rounded, of varying sizes (0.5-2 cm), firm, mobile, painless and covered by normal skin, without inflammatory characteristics. They are located on extensor tendon prominences and are more easily perceived by palpation than by inspection (SZCZYGIELSKA et.al, 2018).

According to Muller et.al (2008), nodules occur preferentially on the elbows, wrists, knees, ankles, occipital region, Achilles tendon and spine. The onset is late (one to two weeks after the other manifestations), regresses rapidly with the start of carditis treatment and rarely persists for more than a month. The nodules are not pathognomonic of RF, as similar structures can be found in other rheumatic diseases, such as polyarticular juvenile idiopathic arthritis, systemic lupus erythematosus and mixed connective tissue disease.

Minor criteria

Castro (2012) describes that the minor signs include non-specific clinical and laboratory characteristics which, together with the major manifestations and evidence of previous streptococcal infection, help to establish the diagnosis of RF.

Arthralgia

According to Culliford - Semmens et.al (2019), isolated arthralgia affects the large joints and is characterized by the absence of functional disability, the presence of which distinguishes it from arthritis. The presence of arthralgia with a migratory and asymmetrical polyarticular pattern involving large joints is highly suggestive of rheumatic fever and is often associated with carditis. The presence of arthritis as a major criterion does not allow arthralgia to be included as a minor criterion.

Fever

According to Mirabel et.al (2015), fever is frequent at the beginning of the acute attack and occurs in almost all arthritis attacks. It has no characteristic pattern. It usually subsides spontaneously within a few days and responds quickly to non-hormonal anti-inflammatory drugs. Patients with carditis not associated with arthritis may have a low-grade fever, while those with pure chorea are afebrile.

PR interval

According to Muller et al (2008), the PR interval on the electrocardiogram may be increased in patients with rheumatic fever, even in the absence of carditis, just as it is in normal individuals. The electrocardiogram should be ordered in all patients with suspected RF and repeated to record the return to normal. In children, the PR interval is considered to be increased when it is above 0.18 s and, in adolescents and adults, above 0.20 s (SZCZYGIELSKA et.al, 2018).

Acute phase reactants

According to Culliford - Semmens et.al (2019), tests of inflammatory activity or acute phase reactants are not specific to RP, but they help to monitor the presence of an inflammatory process (acute phase) and its remission.

According to Mirabel et al (2015), the erythrocyte sedimentation rate (ESR) rises in the first few weeks of the disease. It should be noted that in the presence of anemia, ESR may be overestimated, as well as underestimated in patients with heart failure.

In agreement with De Castro (2012), C-reactive protein (CRP) rises at the beginning of the acute phase and its values decrease at the end of the second or third week. Whenever possible, it should be titrated, as it is more reliable than ESR.

According to Mirabel et.al (2015), acid alpha-1-glycoprotein shows high titers in the acute phase of the disease and remains high for a longer time. It should be used to monitor RF activity.

In protein electrophoresis, alpha-2-globulin is elevated early in the acute phase and can also be used to monitor disease activity (SZCZYGIELSKA et.al, 2018).

According to Muller et.al (2008) it is important to emphasize that any combination of acute phase laboratory tests should be considered as only a minor manifestation of RF.

Complementary tests to assess cardiac involvement in RF

Chest X-ray and electrocardiogram

Ralpf et.al (2021) describes that a chest X-ray is recommended to investigate cardiomegaly and signs of pulmonary congestion. Sequential analysis characterizing an increase in the cardiac area speaks in favour of rheumatic activity, and the presence of pulmonary congestion characterizes severe carditis.

According to Culliford - Semmens et.al (2019), with regard to the electrocardiogram, the findings are non-specific, usually transient and mainly represented by sinus tachycardia, conduction disturbances, ST-T changes and low voltage of the QRS complex and T wave in the frontal plane. A normal electrocardiogram does not exclude cardiac involvement, and the diagnosis of carditis should not be based solely on electrocardiographic abnormalities.

As described above, an increased PR interval is considered a minor criterion. It can be found in patients with RF with or without carditis and can also be observed in normal children. Third-degree AV block and left bundle branch block are rare in active rheumatic disease (SZCZYGIELSKA et.al, 2018).

According to Mirabel et.al (2015), there is a tendency for the QT interval to lengthen in children and adolescents with carditis. An increase in QT dispersion has also been recorded, as well as its reduction as symptoms improve. Occasional cardiac arrhythmias are generally self-limiting and benign. Complex arrhythmias, including torsades de pointes, are rare.

Echocardiography

Recent studies using echocardiographic findings point to a higher prevalence of carditis when compared to estimates based solely on clinical investigation (SZCZYGIELSKA et.al, 2018).

According to Mirabel et.al (2015), persistence or worsening of valve lesions on echocardiography may occur, even with regression of clinical findings.

According to Mirabel et.al (2015), the WHO therefore recommends the use of echocardiography to diagnose subclinical carditis in areas where the disease is endemic (I-B). The echocardiographic criteria for diagnosing subclinical valvitis are described.

According to Muller et al (2008), in the acute phase of the disease, mitral regurgitation is the most frequent alteration. In order to increase the specificity of the echocardiographic diagnosis of mitral regurgitation, morphological valve alterations must also be present.

According to Mirabel et al (2015), mitral regurgitation is caused by ventricular dilatation, mitral annulus dilatation and mitral prolapse. Chordae rupture involving the anterior leaflet can occur exceptionally.

According to Culliford - Semmens et.al (2019), aortic regurgitation is the second most frequent lesion, and obstructive valve lesions do not occur in the initial episodes of RF. Valve thickening is frequent, and focal valve nodules have been observed that disappear during evolution. The diagnosis of valvitis should not be based on isolated lesions on the right side of the heart.

Left ventricular function is normal in the initial episode and, even in recurrences, most patients maintain preserved function. Pericardial effusion, usually small, occurs frequently, but is not always related to the presence of pericardial friction (SZCZYGIELSKA et.al, 2018).

Other diagnostic tests

According to Mirabel et.al (2015), the low sensitivity of endomyocardial biopsy for the diagnosis of carditis limits the use of this technique in clinical investigation. Gallium-67 scintigraphy shows better results when compared to antimyosin scintigraphy, but experience with both methods for imaging myocardial inflammation is limited.

Treatment of acute rheumatic fever

Ralpf et.al (2021) describes that the aim of treating acute RF is to suppress the inflammatory process, minimizing the clinical repercussions on the heart, joints and central nervous system, as well as eradicating EBGA from the oropharynx and promoting relief of the main symptoms.

Other diagnostic tests

According to Mirabel et.al (2015), the low sensitivity of endomyocardial biopsy for the diagnosis of carditis limits the use of this technique in clinical investigation. Gallium-67 scintigraphy shows better results when compared to antimyosin scintigraphy, but experience with both methods for imaging myocardial inflammation is limited.

Treatment of acute rheumatic fever

Ralpf et.al (2021) describes that the aim of treating acute RF is to suppress the inflammatory process, minimizing the clinical repercussions on the heart, joints and central nervous system, as well as eradicating EBGA from the oropharynx and promoting relief of the main symptoms.

General measures

According to Mirabel et al (2015), hospitalization should occur in patients with suspected RF, first outbreak or recurrence, and the need for hospitalization varies according to the severity of the clinical presentation. Hospitalization is indicated for cases of moderate or severe carditis, disabling arthritis and severe chorea. Hospitalization can also be aimed at shortening the time between clinical suspicion and diagnosis, as well as starting treatment quickly. It should also be an opportunity to educate the patient and their family about the disease and the need to adhere to secondary prophylaxis. The length of hospitalization will depend on the control of symptoms, especially carditis.

For many years, bed rest has been recommended, especially before the advent of penicillin. Some studies have shown that bed rest was associated with a reduction in the duration and intensity of carditis, as well as a reduction in the duration and intensity of carditis as well as reducing the duration of the acute outbreak. However, to date, there is no evidence from randomized studies to prove the benefits of this approach (MÜLLER, et al. 2008).

According to Mirabel et al (2015), absolute bed rest is no longer recommended for most patients with RP. Patients with acute RF should, however, be on relative bed rest (at home or in hospital) for an initial period of two weeks. In cases of moderate or severe carditis, relative bed rest should be recommended for a period of 4 weeks. The return to normal activities should be gradual, depending on the improvement of symptoms and the normalization or marked reduction of inflammatory activity tests (ESR and CRP).

Temperature control

Mirabel et.al (2015), describe that if the fever is low, there is no need to administer antipyretic drugs. In cases of higher fever (equal to or greater than 37.8 °C), it is recommended to use paracetamol as the first option, or dipyron as the second option. The use of non-steroidal anti-inflammatory drugs, including acetylsalicylic acid, is not recommended until the diagnosis of RF is confirmed.

Streptococcus eradication

The treatment of pharyngotonsillitis and the eradication of streptococcus from the oropharynx should be carried out when there is clinical suspicion of RF, regardless of the result of the oropharyngeal culture. The aim is to reduce the patient's antigenic exposure to streptococcus and prevent the spread of rheumatogenic strains in the community. In cases of the first outbreak, treatment corresponds to the start of secondary prophylaxis. The therapeutic regimens for eradicating streptococcus are detailed in the Prophylaxis subheading (MÜLLER, et al. 2008).

Treatment of arthritis

According to Joseph et.al (2013), in general, the use of non-steroidal anti-inflammatory drugs (NSAIDs) shows good results in controlling arthritis, leading to the disappearance of signs and symptoms within 24-48 hours. Acetylsalicylic acid (ASA) has remained the first choice for treating joint involvement for over 50 years. In children, the dose initially used is 80-100 mg/kg/day, divided into 4 daily doses. This dose should be reduced to 60 mg/kg/day after two weeks of treatment, if signs and symptoms have improved, and should be maintained for a period of around 4 weeks, in order to cover the period of inflammatory activity of the disease. (I-A) In adults, the recommended dose is 6-8 g/day.

According to Mirabel et.al (2015), in the presence of an acute viral process, it is suggested that ASA be suspended due to the risk of Reye's syndrome.

Joseph et.al (2013) mentions that naproxen is considered a good alternative to ASA, with the same efficacy, easier dosage and better tolerance. The dose used is 10-20 mg/kg/day, taken twice a day, with a treatment duration similar to that of ASA.

Post-streptococcal reactive arthritis may not respond well to treatment with ASA and naproxen. In these cases, the use of indomethacin is indicated (MÜLLER, et al. 2008).

FINAL CONSIDERATIONS

Although it is a relatively easily preventable disease that depends solely on the appropriate treatment of tonsillitis, rheumatic fever, once established with severe cardiac damage.

RF can lead the patient to repeated hospitalizations, complex cardiovascular surgery and drug treatment that is difficult to manage, such as the use of anticoagulants for the rest of their lives, influencing the working capacity of patients and their guardians, and leading to high social costs directly or indirectly, so a simple "sore throat", when treated inappropriately, can lead to serious heart disease, and even death.

Early diagnosis of cases of bacterial tonsillitis among school-age children and referral of these suspected cases for the most appropriate treatment is of the utmost importance.

REFERENCES

- ASMARE, Melkamu Hunegnaw et al. Rheumatic heart disease screening based on phonocardiogram. **Sensors**, v. 21, n. 19, p. 6558, 2021.
- ATHAYDE, Guilherme Rafael Sant'anna. Efeito da valvuloplastia mitral percutânea sobre a complacência do átrio esquerdo em pacientes com estenose mitral reumática. 2015.
- CABRAL, Juliana Chaves et al. Intervenção educativa para a prevenção da febre e cardiopatia reumáticas: ensaio comunitário em um aglomerado urbano subnormal no nordeste do Brasil. 2019.
- CARAPETIS, Jonathan R. et al. Continued Challenge of Rheumatic Heart Disease. **Glob Heart**, v. 8, p. 185-186, 2013.
- CULLIFORD-SEMMENS, Nicola et al. The World Heart Federation criteria raise the threshold of diagnosis for mild rheumatic heart disease: three reviewers are better than one. **International Journal of Cardiology**, v. 291, p. 112-118, 2019.
- DA COSTA, Joicey Melo et al. Impacto da Etiologia sobre a Função Atrial Esquerda em Pacientes com Refluxo Mitral Importante.
- CASTRO, Marildes Luiza. Estudo da função ventricular direita na estenose mitral reumática. 2012.
- FEITOSA, Helvécio N. et al. Mortalidade materna por cardiopatia. **Revista de saúde pública**, v. 25, n. 6, p. 443-451, 1991.
- GOMES, Nayana Flamini Arantes et al. Progressão da regurgitação mitral na cardiopatia reumática: incidência, fatores associados e impacto nos desfechos clínicos. 2022.
- JOSEPH, Nitin et al. Clinical spectrum of rheumatic fever and rheumatic heart disease: a 10 year experience in an urban area of South India. **North American journal of medical sciences**, v. 5, n. 11, p. 647, 2013.
- LEDOS, Pierre-Henri et al. Prevalence of rheumatic heart disease in young adults from New Caledonia. **Archives of cardiovascular diseases**, v. 108, n. 1, p. 16-22, 2015.
- MARIJON, Eloi et al. Prevalence of rheumatic heart disease detected by echocardiographic screening. **New England Journal of Medicine**, v. 357, n. 5, p. 470-476, 2007.
- MARTINS, Joao Francisco BS; NASCIMENTO, Erickson R.; PAPPAS, Gisele L. Diagnóstico Automático de Cardiopatia Reumática em Exames Ecocardiográficos. In: **Anais Estendidos do XXII Simpósio Brasileiro de Computação Aplicada à Saúde**. SBC, 2022. p. 50-55.
- MIRABEL, Mariana et al. Newly diagnosed rheumatic heart disease among indigenous populations in the Pacific. **Heart**, v. 101, n. 23, p. 1901-1906, 2015.
- MÜLLER, Regina Elizabeth et al. **Estudo longitudinal de pacientes portadores de cardiopatia reumática no Rio de Janeiro**. 2008. Tese de Doutorado. Instituto Fernandes Figueira.
- RALPH, Anna P. et al. The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease. **Medical Journal of Australia**, v. 214, n. 5, p. 220-227, 2021.
- REMENYI, Bo et al. Inter-rater and intra-rater reliability and agreement of echocardiographic diagnosis of rheumatic heart disease using the World Heart Federation evidence-based criteria. **Heart Asia**, v. 11, n. 2, 2019.
- REMENYI, Bo et al. Position statement of the World Heart Federation on the prevention and control of rheumatic heart disease. **Nature Reviews Cardiology**, v. 10, n. 5, p. 284-292, 2013.
- REMENYI, Bo et al. World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease—an evidence-based guideline. **Nature reviews cardiology**, v. 9, n. 5, p. 297-309, 2012.
- ROBERTS, Kathryn V. et al. Rheumatic heart disease in Indigenous children in northern Australia: differences in prevalence and the challenges of screening. **Medical Journal of Australia**, v. 203, n. 5, p. 221-221, 2015.
- SAXENA, Anita et al. Echocardiographic prevalence of rheumatic heart disease in Indian school children using World Heart Federation criteria—A multi site extension of RHEUMATIC study (the e-RHEUMATIC study). **International journal of cardiology**, v. 249, p. 438-442, 2017.
- SCHEEL, Amy et al. Community study to uncover the full spectrum of rheumatic heart disease in Uganda. **Heart**, v. 105, n. 1, p. 60-66, 2019.
- SOUZA, Solange Pires Salomé de. **A repercussão da febre reumática e da cardiopatia reumática na vida de crianças e adolescentes: o movimento entre sentir-se saudável e sentir-se doente**. 2006. Tese de Doutorado. Universidade de São Paulo.
- SZCZYGIELSKA, Izabela et al. Rheumatic fever—new diagnostic criteria. **Reumatologia/Rheumatology**, v. 56, n. 1, p. 37-41, 2018.
- VASCONCELOS, Marcelle Cristina da Silva Bastos et al. Fatores associados ao Acidente Vascular Cerebral Isquêmico na Cardiopatia Reumática Crônica. 2020.

WATKINS, David A. et al. Rheumatic heart disease worldwide: JACC scientific expert panel. **Journal of the American College of Cardiology**, v. 72, n. 12, p. 1397-1416, 2018.

WEBB, Rachel; WILSON, N.; LENNON, Diana. Rheumatic heart disease detected by echocardiographic screening. 2007.

WHITE, Harvey et al. Rheumatic heart disease in indigenous populations. **Heart, Lung and Circulation**, v. 19, n. 5-6, p. 273-281, 2010.

ZAHARI, Norazah et al. Pediatric rheumatic heart disease in a middle-income country: A population-based study. **Journal of Tropical Pediatrics**, v. 68, n. 1, p. fmac005, 2022.

ZÜHLKE, Liesl J. et al. The second rheumatic heart disease forum report. **Global heart**, v. 8, n. 3, p. 253-261, 2013.