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REVIEW ARTICLE

Mitral and Aortic Regurgitation in Rheumatic Heart Diseases

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Abstract:

Rheumatic heart disease is a systemic immune process that is sequelae to a beta-hemolytic streptococcal infection of the pharynx. It is most common in developing countries. It is responsible for 250,000 deaths in young people worldwide each year. Over 15 million people have evidence of rheumatic heart disease. The long-term damage to cardiac valves caused by ARF, which can result from a single severe episode or from multiple recurrent episodes of the illness, is known as rheumatic heart disease (RHD) and is a notable cause of morbidity and mortality in resource-poor settings around the world. Although our understanding of disease pathogenesis has advanced in recent years, this has not led to dramatic improvements in diagnostic approaches, which are still reliant on clinical features using the Jones Criteria, or treatment practices. Indeed, penicillin has been the mainstay of treatment for decades and there is no other treatment that has been proven to alter the likelihood or the severity of RHD after an episode of ARF. Recent advances — including the use of echocardiographic diagnosis in those with ARF and in screening for early detection of RHD, progress in developing group A streptococcal vaccines and an increased focus on the lived experience of those with RHD and the need to improve quality of life — give cause for optimism that progress will be made in coming years against this neglected disease that affects populations around the world, but is a particular issue for those living in poverty.The acute illness can be severe, with disabling pain from arthritis, breathlessness and oedema from heart failure, high fevers and choreiform movements that impair activities of daily living. ARF is usually best managed in hospital, often for a 2–3 week period, by which time the diagnosis is confirmed and the symptoms are treated. Although most of the clinical features of ARF will resolve during this short hospital stay, the cardiac valvular damage might persist. This chronic valvular damage is known as rheumatic heart disease (RHD) and is the major cause of morbidity and mortality from ARF. ARF can recur as a result of subsequent GAS infections and each recurrence can worsen RHD. Thus, the priority in disease management is to prevent ARF recurrences using long-term penicillin treatment, which is known as secondary prophylaxis.

The persistent damage to heart valves resulting in mitral and/or aortic regurgitation, or in long-standing cases stenosis, that remains as a result of acute rheumatic fever with rheumatic carditis. Complications of rheumatic heart disease include heart failure, embolic stroke, endocarditis and atrial fibrillation.

Keywords: ***atrial fibrillation, mitral stenosis, left atrial, mitral and aortic regurgitation, rheumatic heart disease.***

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Introduction:

Background: Unlike the Western world, valvular disease ranks among the major cardiovascular afflictions in Bangladesh. Acute rheumatic fever and chronic rheumatic valvular disease in their most virulent form are still commonly encountered and impose a huge burden on limited healthcare resources

Epidemiology of Streptococcal Pharyngitis:

Although neither streptococci nor their products can be found in the heart of patients with acute rheumatic fever or chronic rheumatic heart disease and no animal model exists, there is sufficient circumstantial evidence to implicate antecedent pharyngitis due to group A

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streptococci in the pathogenesis of this disease. It is intriguing to note, however, that although there has been no apparent or documented decline in group A streptococci pharyngitis, the incidence of acute rheumatic fever in industrialized countries has dropped precipitously to below 1/100 000, whereas in Sudan it exceeds 100/100 000. Although these differences may in part reflect less overcrowding and more effective antibiotic treatment of group A streptococci pharyngitis in Western countries, differences both in the frequency of infection with rheumatogenic M-protein serotypes and in their virulence may be important.[10]

The incidence of initial cases of ARF is highest in children aged 5–14 years, although first episodes do occur in younger children, with reported cases of ARF in those as young as 2–3 years old. Initial episodes can also occur in older adolescents and adults, although cases in people >30 years of age are rare. By contrast, recurrent episodes often affect slightly older children, adolescents and young adults but are rarely observed beyond the age of 35–40 years.[1]

Osteoporosis is a chronic disease characterized by an increased risk of fragility fracture. Patients affected by rheumatic diseases are at greater risk of developing osteoporosis. The purpose of the present review is to discuss the pathogenesis, epidemiology, and treatment of osteoporosis in patients affected by rheumatic diseases with special focus for rheumatoid arthritis, psoriatic arthritis, spondyloarthritis, systemic lupus erythematosus, systemic sclerosis, vasculitides, Sjogren syndrome, and crystal-induced arthritis.[2]

Patients with systemic autoimmune rheumatic diseases, particularly RA, SLE, SS and idiopathic inflammatory myopathies, are at increased risk of developing malignancies. Cancer occurrence adds to the disease burden in these patients, adversely affecting quality of life and life expectancy. This risk is related to the pathobiology of the underlying rheumatic disease including the inflammatory burden, immunological defects, and personal and environmental exposure such as smoking and some viral infections.[3]

A disease of the poor, RHD is one of the most neglected diseases. Several challenges are unique to the acute rheumatic fever/RHD continuum and contribute to its persistence, including its sequestration among the poorest, its protracted natural history, the erratic availability of penicillin, and the lack of a concerted effort in endemic regions. However, there is cause for optimism following a resurgence in scientific interest over the last 15 years. This review presents the latest advancements in epidemiology, diagnosis, and management. It also discusses pressing research questions on disease pathophysiology, the barriers to implementation of effective management strategies, and pragmatic policy solutions required for translation of current knowledge into meaningful action.[4]

Rheumatic heart disease (RHD), a sequela of acute rheumatic fever (ARF), affects 40.5 million people worldwide. The burden of disease disproportionately falls on low- and middle-income countries (LMIC) and sub-populations within high-income countries (HIC). Advances have been made in earlier detection of RHD, though several barriers to ideal management persist Expert.

Systolic AR is a unique hemodynamic phenomenon in patients with acute rheumatic carditis involving both mitral and aortic valves and occurs in presence of severe MR.[5]

The prevalence of RHD has been rising steadily since 1990, reaching 40.5 million in 2019, and accounting for 306,000 deaths annually as a consequence of severe valvular disease. Mitral regurgitation (MR) is the most common valvular abnormality at the early RHD stages, usually associated with ongoing inflammatory rheumatic activity in children.This pure MR may resolve with effective treatment of the acute carditis and continued prophylactic therapy.[6]

Acute rheumatic fever (ARF) with carditis can lead to the development of rheumatic heart disease in children and young adults.

Children diagnosed with rheumatic carditis during 2005-2020 at Siriraj Hospital (Bangkok, Thailand) were retrospectively enrolled. Trivial, and mild regurgitation were grouped as non-clinically significant (NCS) regurgitation. Valvular regression was defined moderate-severe regurgitation improving to NCS regurgitation.Thai children with rheumatic carditis had a high incidence of valvular regurgitation; however, the valvular damage was improved in most patients. High ESR and severe carditis independently predict MR improvement.[7]

Discussion:

What causes rheumatic heart disease? Rheumatic heart disease is caused by rheumatic fever. This is an inflammatory disease that can affect many connective tissues, especially in the heart, joints, skin, or brain. The heart valves can be inflamed and become scarred over time but our article discussed how rheumatic fever developed the carditis

Global burden: A 2005 systematic review, carefully designed to avoid over-estimating the disease burden of GAS infection, concluded that there were approximately 471,000 cases of ARF each year (336,000 in children aged 5–14 years), 15.6–19.6 million prevalent cases of RHD and approximately 350,000 annual deaths as a result of ARF or RHD; almost all deaths occurred in low-income and middle-income countries.[1]

The clinical features of 205 cases of rheumatic heart disease in Bangladesh, including unique two-dimensional echocardiographic findings, were reported, and these were compared with those of 387 Japanese cases. The percentage of mitral stenosis (MS), aortic valvular diseases including both aortic stenosis and aortic regurgitation (A), mitral stenosis with aortic valvular diseases (MS + A) were almost the same between the two countries, but that of mitral regurgitation (MR) was higher, mitral stenosis and regurgitation (MSR) was lower in Bangladesh.[8]

The most important determinant of marked aggravation of MR is the occurrence of a new flail leaflet followed by an increase in annular diameter, which results in reduced leaflet coaptation. Another study evaluating patients with MV prolapse demonstrated that only mitral annular diameter is a predictor of progression to severe MR.[9]

In patients with rheumatic MV disease, a chronic pressure-volume overload on the left atrium leads to a range of adaptive processes that include LA remodelling, which encompasses changes in atrial size, function, and shape. LA enlargement also reflects the intrinsic compliance of the left atrium, risk of subsequent AF, and overall disease severity. In the presence of mixed MV disease, LA is affected by both stenosis and regurgitation, which aggravates its remodeling over time with the progression of MR as a consequence of the mitral annulus size. Subsequent progression of primary rheumatic lesions should also be considered. Turbulent flow drives valvular tissue injury, continuously stimulating inflammatory processes and mechanical trauma, which contribute to perpetuate the valvular damage. Additionally, patients with RHD often have associated AF, which may contribute to the progression of LA and annular dilation thus increasing the severity of MR

In patients with RHD with a full spectrum of MR severity, progression of MR occurs over time predicted by age and LA volume, corrected by competing risks. LA enlargement may play a role in the link between primary MR and secondary MR in patients with RHD. Assessment of MR progression may provide important insight into the long-term consequences of the disease and the rationale for patient management.[6]

Conclusion:

In patients with RHD with a full spectrum of MR severity, progression of MR occurs over time predicted by age and LA volume, corrected by competing risks. LA enlargement may play a role in the link between primary MR and secondary MR in patients with RHD. Assessment of MR progression may provide important insight into the long-term consequences of the disease and the rationale for patient management.

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