

Recommendations

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Recommendations

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I. INTRODUCTION

Acute rheumatic fever (ARF) and its chronic sequela, rheumatic heart disease (RHD), are major health problems globally.¹ Worldwide, RHD is one of the most common cardiovascular diseases in children and young people under 25 years of age and remains an important cause of morbidity and mortality throughout developing countries.¹ RHD is the single most important cause of cardiac valve disease in many parts of the world. With ongoing emigration and immigration in recent decades, RHD has crossed borders and become a global burden.

While minimum echocardiographic criteria for the diagnosis of RHD and diagnostic criteria for ARF incorporating Doppler echocardiography have been published previously,^{2,3} a guide to comprehensive noninvasive evaluation of rheumatic cardiac lesions is lacking. The purpose of this document is to provide recommendations for the use of echocardiography in the diagnosis, classification, and risk assessment of RHD and to help educate clinicians and sonographers worldwide on the essential role of echocardiography in the comprehensive assessment of the disease, and to provide therapeutic directions.

II. EPIDEMIOLOGY OF RHEUMATIC HEART DISEASE

A recent report identified more than 33 million cases of RHD globally.⁴ However, these statistics may under-represent the disease burden, owing to under-reporting, scarce healthcare resources, and few systematic registry programs in the low-income countries where the disease is endemic.¹ Adding echocardiography to the screening protocols leads to a much higher estimate of RHD prevalence, up to ten times more than identified by clinical screening alone.⁵ The highest prevalence is found in Oceania, South Asia, and central sub-Saharan Africa.⁴

While the incidence is low in industrialized nations, ARF and RHD are not uncommon among certain indigenous populations within these countries.^{6,7} In addition, new cases of RHD are being seen in industrialized nations with demographic shifts due to immigration from underdeveloped countries.^{1,6} In the United States, RHD ranks 8th among the cardiovascular causes in terms of deaths, years of life lost, and the age-standardized mortality rate at the national level.⁸

Given the overall disease burden and the high likelihood of developing RHD from the initial episode of ARF, clinicians must make an accurate diagnosis and promptly institute appropriate antibiotic therapy as well as prophylaxis. With the recent incorporation of echocardiographic findings into the diagnostic criteria for both ARF and RHD, more subclinical cases of rheumatic carditis and RHD are being recognized.^{2,3} The application of echocardiography, therefore, has a fundamental implication on modifying the worldwide burden of rheumatic carditis and RHD, its treatment, and prognosis in this globalized era.

III. BASIC CONCEPTS OF PATHOPHYSIOLOGY, CLINICAL PRESENTATION, AND SCREENING

A. Pathophysiology and Clinical Presentation

RHD is the long-term consequence of immune-mediated injury to the heart and cardiac valves following infection by group A β -hemolytic streptococcus (GAS), also known as *Streptococcus pyogenes* (Fig. 1A). ARF is an acute illness following the infection and usually manifests 2-4 weeks after GAS tonsillopharyngitis. Clinical presentations of ARF can be variable and may include fever, arthritis, carditis, chorea, subcutaneous nodules, and erythema marginatum. Of these, carditis is the most common presentation (50%-70%) during the first episode of ARF.³ While acute rheumatic carditis can present as pancarditis involving the endocardium, myocardium, and pericardium, it almost always involves valvulitis, inflammation of the valvular endocardium. Subclinical carditis is defined by the presence of echocardiographically-detected valvulitis without clinical signs of carditis, and may be present in up to 53% of all cases of ARF.⁹ The pathogenesis of acute rheumatic carditis is thought to be immune-mediated following GAS infection. Antibodies developed in response to GAS pharyngitis cross-react with cardiac proteins through a process termed 'molecular mimicry'.¹⁰ The subsequent autoimmune response causes the major manifestations of ARF. The presence of ARF in only a subset of children untreated for streptococcal pharyngitis and the fact that only a minority of the affected children develop RHD also suggest a possible role of host susceptibility.¹¹ Aschoff bodies (Fig. 1B), the pathognomonic lesions of the rheumatic process that form in the endocardium of the heart valve and in the myocardium, are thought to be sites of granulomatous inflammation as a result of T lymphocyte-mediated delayed hypersensitivity against GAS antigens.¹² Damage to cardiac valves may be progressive, ultimately resulting in RHD, the chronic sequela of carditis, with the development of valvular stenosis and regurgitation, pulmonary hypertension (PH), and heart failure. Thus, the immune-mediated injury responsible for acute rheumatic carditis is dependent mainly on a triad of factors: infection with GAS, a susceptible host, and an aberrant host response.

B. Echocardiographic Screening in Acute Rheumatic Fever

Echocardiography significantly enhances the diagnostic yield of carditis in suspected cases because mild and moderate pathological valvular regurgitations may be missed with auscultation alone. This is important because a longer duration of secondary prophylaxis is recommended to prevent the recurrence of rheumatic fever in patients with valvulitis.¹³ In 2015, the use of echocardiography was formally incorporated into the revised Jones criteria as a tool to diagnose rheumatic carditis, with subclinical carditis (or echocardiographic valvulitis) recognized as a major criterion for diagnosis of ARF.³ Handheld point of care cardiac ultrasound (cardiac POCUS)