

Acute rheumatic fever (ARF) is a multisystem disease resulting from an autoimmune reaction to infection with group A *Streptococcus*. Although many parts of the body may be affected, almost all of the manifestations resolve completely. The major exception is cardiac valvular damage (rheumatic heart disease [RHD]), which may persist after the other features have disappeared.

GLOBAL CONSIDERATIONS

ARF and RHD are diseases of poverty. They were common in all countries until the early twentieth century, when their incidence began to decline in industrialized nations. This decline was largely attributable to improved living conditions—particularly less crowded housing and better hygiene—which resulted in reduced transmission of group A streptococci. The introduction of antibiotics and improved systems of medical care had a supplemental effect.

The virtual disappearance of ARF and reduction in the incidence of RHD in industrialized countries during the first half of the twentieth century unfortunately was not replicated in developing countries, where these diseases continue unabated. RHD is the most common cause of acquired heart disease in children in developing countries and is a major cause of mortality and morbidity in adults as well. It has been estimated that between 29.7 and 43.1 million people worldwide are affected by RHD, with >300,000 deaths occurring each year. Some 95% of ARF cases and RHD deaths now occur in developing countries, with particularly high rates in sub-Saharan Africa, Pacific nations, Australasia, and South and Central Asia. The pathogenetic pathway from exposure to group A *Streptococcus* followed by pharyngeal or superficial skin infection and subsequent development of ARF, ARF recurrences, and development of RHD and its complications is associated with a range of risk factors and,