ARTICLE

Acute Rheumatic Fever: Global Persistence of a Preventable Disease

Francine Bono-Neri, MA, RN, PNP

ABSTRACT

The persistence of acute rheumatic fever continues to be seen globally. Once thought to be eradicated in various parts of the world, the disease came back with a vengeance secondary to a lack of diligence on the part of providers. Today, the global burden of group A streptococcal infection, the culprit of the numerous sequelae manifested in acute rheumatic fever, is considerable. Although a completely preventable disease, rheumatic fever continues to exist. It is a devastating disease that involves long-term, multisystem treatment and monitoring for patients who were unsuccessful at eradicating the precipitating group A streptococcal infection. Prevention is the key to resolving the dilemma of the disease's global burden, yet the method to yield its prevention still remains unknown. Thus, meticulous attention to implementing proper treatment is the mainstay and remains a top priority. J Pediatr Health Care. (2016) ■, ■-■.

KEY WORDS

Acute rheumatic fever (ARF), carditis, group A *Streptococcus* (GAS), Jones criteria, pharyngitis

Despite the efficacy of antibiotics against group A *Streptococcus* species in reducing the incidence of acute rheumatic fever (ARF), the global burden and chronic sequelae of the disease continue to exist. Although North America and Europe have seen a reduced frequency in acute rheumatic fever over the past several decades (Bach, 2015), providers must not exclude this diagnosis from their differential. Notwithstanding accessibility to antimicrobials, countries continue to see ARF as a major cause of serious valvular heart disease (Parnaby & Carapetis, 2010). Research and new guidelines continue to evolve because of its persistent prevalence.

In the United States, a decline in ARF was seen in the 30 years after World War II. The annual occurrence dropped by more than 90%, which was believed to reflect ameliorated living conditions, overall improved hygiene, and the use of antibiotics (Congeni, 1992). By the early 1980s, ARF had reached such an all-time low in the United States that some providers began to question their ardency in treating streptococcal pharyngitis. It was not until the major epidemic at the beginning of 1984 that a resurgence was witnessed in various regions of the United States (Congeni, Rizzo, Congeni, & Sreenivasan, 1987; Hosier, Craenen, Teske, & Wheller, 1987; Veasy et al., 1987; Wald, Dashefsky, Fedit, Chiponis, & Byers, 1987). Unlike the traditional outbreaks found in crowded and impoverished inner-city ghettos, these cases occurred primarily in children of White, middle-class families, many of whom resided in suburban or rural environments (Congeni, 1992). In the present day, although isolated cases of ARF continue to be seen in modernized countries, most are found in countries with limited resources and in poorly represented aboriginal groups (World Health Organization [WHO], 2005). Various regions of South America, the Middle East, India, and Africa are showing particular risk of ARF for children (Casey, Solomon, Gaziano, Miller, & Loscalzo, 2013; Tibazarwa, Volmink, & Mayosi, 2008).

Global burden of disease, up until the early 1990s, was assessed using a narrow criterion. Global studies of

Francine Bono-Neri, Adjunct Clinical Instructor, The Barbara H. Hagan School of Nursing, Molloy College, Rockville Centre, NY.

Conflicts of interest: None to report.

Correspondence: Francine Bono-Neri, MA, RN, PNP, The Barbara H. Hagan School of Nursing, Molloy College, 1000 Hempstead Ave, Rockville Centre, NY 11571; e-mail: fbono-neri@molloy.edu.

0891-5245/$36.00

http://dx.doi.org/10.1016/j.pedhc.2016.09.001

population mortality were the main focus. These studies failed to consider morbidity that arose from disorders and injuries that were not fatal but nevertheless affected a person's functioning in an adverse manner (Degenhardt, Whiteford, & Hall, 2014). Measuring the impact of disease and its global burden was radically transformed in 1993, when estimates of causes of global disease burden used a new summary measure. This new measure, known as Disability-Adjusted Life Years, "simultaneously accounted for both premature mortality and the prevalence, duration and severity of the non-fatal consequences of disease and injury" (Lopez, 2005, Abstract, para. 1). The World Health Report (World Health Organization, 2005) states that approximately 18.1 million people were suffering from a serious group A *Streptococcus* (GAS) disease and that approximately 1.78 million new cases were being seen annually as of 2005. In addition, the report also states that GAS infections were identified as the ninth leading cause of worldwide mortality from a single pathogen. Global burden, defined by the WHO as those afflicted, found an overall burden of 471,000 annual cases of ARF (Zühlke & Steer, 2013). Although prompt diagnosis and treatment are imperative, prevention is pivotal to success.

WHAT IS ARF?

ARF is a nonsuppurative sequela that manifests 2 to 4 weeks after untreated pharyngitis and is caused by GAS (Casey et al., 2013). Some speculate that in various resource-limited tropical regions, ARF has been seen after pyoderma (Parks, Smeesters, & Steer, 2012); however, for the purpose of this article, only GAS pharyngitis will be addressed. Its predisposition toward inflammation of connective tissue classifies ARF as a collagen disease. The heart, joints, subcutaneous tissue, and central nervous system are involved. When involvement of the heart is present, "damage to affected cardiac valves may be chronic and progressive" (Gibofsky, 2016, para. 1), yielding deterioration in cardiac function. Although the exact pathogenesis of ARF remains unclear, antibodies produced after GAS infection react with the body's cells and produce characteristic lesions in target areas. The major benefits of adequate treatment for GAS pharyngitis is the prevention of ARF and reduction of communicability.

Before examining the symptomatology of ARF, certain key points need to be addressed. First, acute rheumatic fever occurs after an untreated GAS pharyngitis (Beaudoin et al., 2015). Second, the prevention of ARF can be accomplished with the use of appropriate antibiotic treatment up to 9 days after the onset of pharyngitis (Watson, Jallow, Doare, Pushparajah, & Anderson, 2015). Third, the peak incidence of pharyngitis caused by GAS is in children ages 5 to 15 years (Wessels, 2011) and accounts for 20% to 30% or more of acute pharyngitis cases in this population (Shulman et al., 2012).

Diagnosis of GAS pharyngitis is paramount to preventing future sequelae. Findings highly suggestive of a viral pharyngitis include conjunctivitis, coryza, cough, hoarseness, anterior stomatitis, and/or diarrhea (Shulman et al., 2012). Clinical signs suggestive of GAS pharyngitis include fever, headache, palatal petechiae, patchy tonsillopharyngeal exudates, anterior cervical lymphadenitis, scarlatiniform rash, nausea, vomiting, abdominal pain, and history of sudden onset of sore throat. Epidemiologic findings include an age of 5 to 15 years old, history of ill contacts with GAS pharyngitis, prevalence in the community, and seasonal timing of winter and early spring for areas with temperate climates (Gerber et al., 2009). These findings are highly indicative of the diagnosis, yet the criterion standard for confirmation to determine positive GAS colonization is by the rapid antigen detection test and/or culture (Shulman et al., 2012). Although a rudimentary test, it must be correctly executed by vigorously swabbing both tonsils and the posterior pharynx (Gerber et al., 2009). With most rapid antigen detection tests, a negative result does not rule out the presence of GAS; therefore, a throat culture should be performed as well (Gerber et al., 2009). If the results are positive for GAS, treatment must be initiated. Treatment options are as listed in Table 1. Not only is it essential to prescribe the appropriate antibiotic for your patient, but it is also crucial to find one that will elicit total compliance to ensure eradication. Hence, some practitioners will use a once-daily dosing such as azithromycin or a one-time intramuscular injection where compliance would have posed as a major concern. It is essential that providers educate patients and parents on complete treatment adherence.

> Diagnosis of GAS pharyngitis is paramount to preventing future sequelae.

WHAT IS THE CLINICAL PRESENTATION OF ARF, AND HOW IS IT DIAGNOSED?

On June 16, 1944, the first speaker at the Annual Session of the American Medical Association Symposium on Rheumatic Fever was a physician named T. Duckett Jones. He presented in a revolutionary and clinically documented manner through which he outlined the diagnostic criteria for rheumatic fever. Historically, these criteria have depicted the clinical standards to establish the diagnosis of ARF. Publicized in the *Journal of the American Medical Association,* the Jones criteria were augmented by the American Heart Association (AHA) in 1992. At a workshop sponsored by the AHA in 2000, the criteria were reaccredited and remained an invaluable tool in diagnosing ARF. However, because of the advances in imaging studies and the evolving role of echocardiography, a re-examination