

**TABLE 381-4 AMERICAN HEART ASSOCIATION RECOMMENDATIONS FOR DURATION OF SECONDARY PROPHYLAXIS\***

Category of Patient	Duration of Prophylaxis
Rheumatic fever without carditis	For 5 years after the last attack or 21 years of age (whichever is longer)
Rheumatic fever with carditis but no residual valvular disease	For 10 years after the last attack, or 21 years of age (whichever is longer)
Rheumatic fever with persistent valvular disease, evident clinically or on echocardiography	For 10 years after the last attack, or 40 years of age (whichever is longer); sometimes lifelong prophylaxis

\*These are only recommendations and must be modified by individual circumstances as warranted. Note that some organizations recommend a minimum of 10 years of prophylaxis after the most recent episode, or until 21 years of age (whichever is longer), regardless of the presence of carditis with the initial episode.

Source: Adapted from AHA Scientific Statement Prevention of Rheumatic Fever and Diagnosis and Treatment of Acute Streptococcal Pharyngitis. *Circulation* 119:1541, 2009.

(recurrences become less likely with increasing time), age (recurrences are less likely with increasing age), and the severity of RHD (if severe, it may be prudent to avoid even a very small risk of recurrence because of the potentially serious consequences) (Table 381-4). Secondary prophylaxis is best delivered as part of a coordinated RHD control program, based around a registry of patients. Registries improve the ability to follow patients and identify those who default from prophylaxis and to institute strategies to improve adherence.

**VIDEO 381-1A** Transthoracic echocardiographic images of a 9-year-old girl with first episode of acute rheumatic fever. Images demonstrate the typical echocardiographic findings of acute rheumatic carditis. The valve leaflets are relatively thin and highly mobile. The failure of coaptation of the mitral valve leaflets is the result of chordal elongation and annular dilatation. The mitral valve regurgitation is moderate with a typical posterolaterally directed regurgitant jet of rheumatic carditis. **A.** Acute rheumatic carditis (apical four-chamber view echocardiogram).

**VIDEO 381-1B** Transthoracic echocardiographic images of a 9-year-old girl with first episode of acute rheumatic fever. Images demonstrate the typical echocardiographic findings of acute rheumatic carditis. The valve leaflets are relatively thin and highly mobile. The failure of coaptation of the mitral valve leaflets is the result of chordal elongation and annular dilatation. The mitral valve regurgitation is moderate with a typical posterolaterally directed regurgitant jet of rheumatic carditis. **B.** Acute rheumatic carditis (apical four-chamber view color Doppler echocardiogram).

**VIDEO 381-1C** Transthoracic echocardiographic images of a 9-year-old girl with first episode of acute rheumatic fever. Images demonstrate the typical echocardiographic findings of acute rheumatic carditis. The valve leaflets are relatively thin and highly mobile. The failure of coaptation of the mitral valve leaflets is the result of chordal elongation and annular dilatation. The mitral valve regurgitation is moderate with a typical posterolaterally directed regurgitant jet of rheumatic carditis. **C.** Acute rheumatic carditis (parasternal long-axis view echocardiogram).

**VIDEO 381-1D** Transthoracic echocardiographic images of a 9-year-old girl with first episode of acute rheumatic fever. Images demonstrate the typical echocardiographic findings of acute rheumatic carditis. The valve leaflets are relatively thin and highly mobile. The failure of coaptation of the mitral valve leaflets is the result of chordal elongation and annular dilatation. The mitral valve regurgitation is moderate with a typical posterolaterally directed regurgitant jet of rheumatic carditis. **D.** Acute rheumatic carditis (parasternal long-axis view color Doppler echocardiogram).

**VIDEO 381-2A** Transthoracic echocardiographic images are from a 5-year-old boy with chronic rheumatic heart disease with severe mitral valve regurgitation and moderate mitral valve stenosis. Images demonstrate the typical echocardiographic findings in advanced chronic rheumatic heart disease. Both the anterior and posterior mitral valve leaflets are markedly thickened. During diastole, the

motion of the anterior mitral valve leaflet tip is restricted with doming of the body of the leaflet toward the interventricular septum. This appearance is commonly described as a “hockey stick” or an “elbow” deformity. **A.** Chronic rheumatic heart disease (parasternal long-axis view). **VIDEO 381-2B** Transthoracic echocardiographic images are from a 5-year-old boy with chronic rheumatic heart disease with severe mitral valve regurgitation and moderate mitral valve stenosis. Images demonstrate the typical echocardiographic findings in advanced chronic rheumatic heart disease. Both the anterior and posterior mitral valve leaflets are markedly thickened. During diastole, the motion of the anterior mitral valve leaflet tip is restricted with doming of the body of the leaflet toward the interventricular septum. This appearance is commonly described as a “hockey stick” or an “elbow” deformity. **B.** Chronic rheumatic heart disease (apical two-chamber view echocardiogram).

## 382 Systemic Sclerosis (Scleroderma) and Related Disorders

John Varga

### DEFINITION

Systemic sclerosis (SSc) is an uncommon connective tissue disorder characterized by multisystem involvement, heterogeneous clinical manifestations, a chronic and often progressive course, and significant disability and mortality. Multiple genes contribute to disease susceptibility; however, environmental exposures are likely to play a major role in causing SSc. The early stage of the disease is associated with prominent inflammatory features. Over time, functional and structural alterations in multiple vascular beds and progressive visceral organ dysfunction due to fibrosis dominate the clinical picture. Although thickened skin (scleroderma) is the distinguishing hallmark of SSc, skin induration can occur in localized forms of scleroderma and other disorders (Table 382-1). Patients with SSc can be broadly

**TABLE 382-1 CONDITIONS ASSOCIATED WITH SKIN INDURATION**

Systemic sclerosis (SSc)
Limited cutaneous SSc
Diffuse cutaneous SSc
Localized scleroderma
Guttate (plaque) morphea, diffuse (pansclerotic) morphea, bullous morphea
Linear scleroderma, coup de sabre, hemifacial atrophy
Pansclerotic morphea
Overlap syndromes
Mixed connective tissue disease
SSc/polymyositis
Stiff skin syndrome
Diabetic scleredema and scleredema of Buschke
Scleromyxedema (papular mucinosis)
Chronic graft-versus-host disease
Diffuse fasciitis with eosinophilia (Shulman's disease, eosinophilic fasciitis)
Chemically induced and drug-associated scleroderma-like conditions
Vinyl chloride–induced disease
Eosinophilia-myalgia syndrome (associated with L-tryptophan)
Nephrogenic systemic fibrosis (associated with gadolinium)
Paraneoplastic syndrome