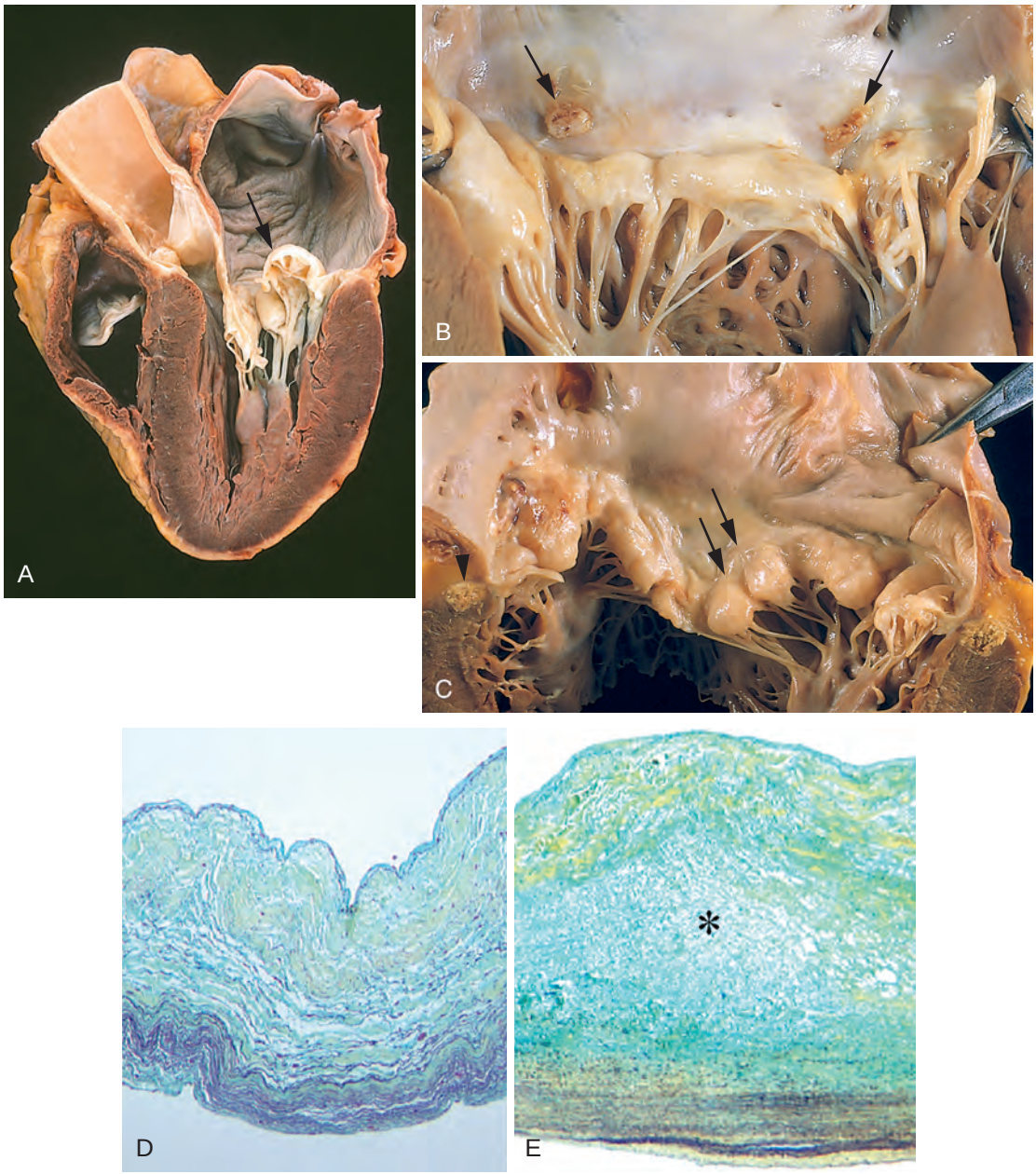

Figure 12-22 Myxomatous degeneration of the mitral valve. **A,** Long axis view (left ventricle is on the *right*) demonstrating hooding with prolapse of the posterior mitral leaflet into the left atrium *(arrow).* **B,** Opened valve, showing pronounced hooding of the posterior mitral leaflet with thrombotic plaques at sites of leaflet-left atrium contact *(arrows).* **C,** Opened valve with pronounced hooding *(double arrows)* in a patient who died suddenly. Note also mitral annular calcification *(arrowhead).* Normal heart valve **(D)** and myxomatous mitral valve **(E).** In myxomatous valves, collagen in the fibrosa is loose and disorganized, proteoglycan deposition *(asterisk)* in the spongiosa is markedly expanded, and elastin in the atrialis is disorganized. (**A,** Courtesy William D. Edwards, MD, Mayo Clinic, Rochester, Minn; **D, E,** Movat pentachrome stain, in which collagen is yellow, elastin is black, and proteoglycans are blue). From Rabkin E, et al: Activated interstitial myofibroblasts express catabolic enzymes and mediate matrix remodeling in myxomatous heart valves. Circulation 104:2525-2532, 2001.)

Rheumatic Fever and Rheumatic Heart Disease

Rheumatic fever (RF) is an acute, immunologically mediated, multisystem inflammatory disease classically occurring a few weeks after an episode of group A streptococcal pharyngitis; occasionally, RF can follow streptococcal infections at other sites, such as the skin. Acute rheumatic carditis is a common manifestation of active RF and may progress over time to chronic rheumatic heart disease (RHD), mainly manifesting as valvular abnormalities.

RHD is characterized principally by deforming fibrotic valvular disease, particularly involving the mitral valve; *indeed, RHD is virtually the only cause of mitral stenosis*. The incidence and mortality rate of RF and RHD have declined remarkably in many parts of the world over the past century, as a result of improved sanitation, and rapid diagnosis and treatment of streptococcal pharyngitis. Nevertheless, in developing countries, and in many crowded, economically depressed urban areas, RHD remains an important public health problem, affecting an estimated 15 million people.