

case of death or serious cardiovascular disease as a direct result of exposure to a beta-blocker.¹ Side effects of beta-blockers are well known; these include transient bradycardia and hypotension that warrant close monitoring at the onset of treatment. Bronchospasm is usually seen as an exacerbation in patients with underlying reactive airways; a family history with regard to atopy or repeated wheezing should be obtained. Beta-blockers decrease lipolysis, glycogenolysis, and gluconeogenesis that predispose patients to hypoglycemia. They also mask some beta-sympathetic-related hypoglycemic symptoms. The first week of life is a critical period when neonates gradually reach their optimal milk intake and spontaneous hypoglycemia is more likely to develop; beta-blockers clearly should be avoided during this period. Most infants treated for an infantile hemangioma are older and have a normal food

intake, and the conditions described in the article by Burns et al.² do not apply.

A large multicenter study is in the planning stage. We hope that this study will lead to elaborate, reasonable, and objective recommendations concerning the use of beta-blockers in this indication.

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Cardiac Amyloidosis with the E526V Mutation of the Fibrinogen A α -Chain

TO THE EDITOR: Proteinuria developed in a 48-year-old man in 2003. His mother had died 10 years earlier from renal amyloidosis. Laboratory tests showed isolated nephrotic-range proteinuria (urinary protein excretion, >7 g per 24 hours) with normal creatinine clearance (estimated glomerular filtration rate with the use of the Modification of Diet in Renal Disease equation, 96 ml per minute per 1.73 m² of body-surface area); no monoclonal gammopathy was found. A specimen from a renal biopsy contained amyloid deposits located exclusively in the mesangium of the glomeruli (Fig. 1A). The hereditary transmission of the disease led us to perform a molecular analysis for hereditary amyloidosis (associated with transthyretin, fibrinogen, apolipoprotein, or lysozyme); only the E526V mutation of the α -chain of fibrinogen A, the most frequent mutation resulting in fibrinogen amyloidosis, was identified.

Three years later, the patient reported dyspnea (New York Heart Association class II), which gradually increased; 4 years later, a cardiac arrhythmia necessitated implantation of a defibril-

lator. The patient had no family history of heart disease. Cardiac ultrasonography suggested cardiac amyloidosis, with septal hypertrophy, a left intraventricular gradient of 40 mm Hg, and filling impairment.¹ Circumferential thickening of the left ventricle was observed on cardiac magnetic resonance imaging, with focal late-phase contrast enhancement of the posteroseptal territory of the midventricular region. Whereas diffuse enhancement is characteristic of AL amyloidosis,² focal enhancement suggests non-AL amyloidosis.³ Specimens from multiple myocardial biopsies showed amyloid deposits in subendocardial and perivascular areas (Fig. 1B), definitively establishing the diagnosis of cardiac amyloidosis. This amyloid material was specifically labeled with anti-fibrinogen antibodies (Dako F0111), whereas the results of staining with antibodies against λ -chain and κ -chain were negative (Fig. 1C).

Since fibrinogen A α -chain amyloidosis was first described in 1993,⁴ about 40 cases have been reported. Renal involvement is always present,

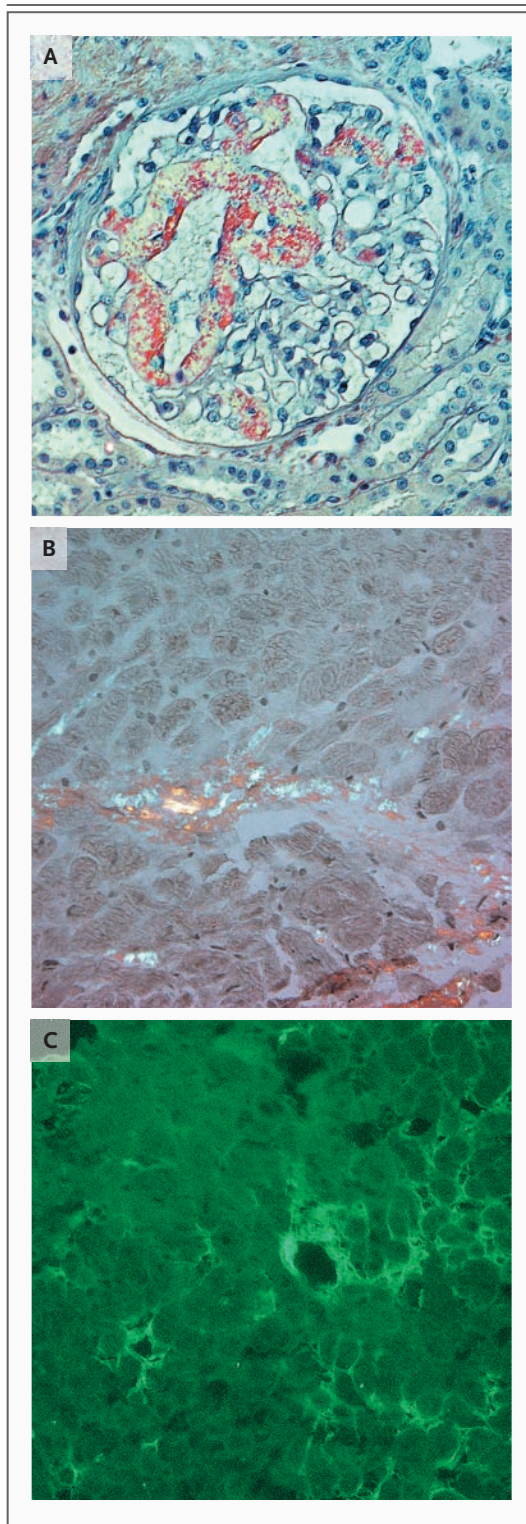


Figure 1. Amyloid Deposits in Specimens from Renal and Myocardial Biopsies.

A renal-biopsy specimen (Panel A) stained with Congo red contains amyloid deposits in the mesangium of the glomeruli, and a myocardial-biopsy specimen (Panel B), also stained with Congo red, contains amyloid deposits in the interstitium and around small vessels. Immunofluorescence staining with antifibrinogen antibodies in a myocardial-biopsy specimen (Panel C) shows that amyloid material has formed a latticework pattern around the myocytes.

and liver involvement, although rarely observed, has generally resulted in severe disease. Splenic deposits are usually asymptomatic and are frequently observed at autopsy.⁵

Our report describes cardiac involvement in fibrinogen A α -chain amyloidosis. The hereditary transmission of the disease, the presence of amyloid deposits in specimens from renal and myocardial biopsies, the specific staining with antifibrinogen antibody, and the presence of the E526V mutation confirm that our patient had fibrinogen A α -chain amyloidosis with both renal and cardiac involvement. This case shows that fibrinogen A α -chain amyloidosis may involve the heart, leading to life-threatening complications.

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