Restrictive Cardiomyopathy in Familial Amyloidosis TTR-Arg-50
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To the Editor:

The report by Singer et al is elegantly illustrated and provides the practicing clinician with an up-to-date flow chart on how to diagnose amyloidosis as the cause of restrictive cardiomyopathy. However, it is disappointing that the authors did not stress the fact that familial amyloidotic polyneuropathy is an autosomal dominant hereditary form of amyloidosis for which effective treatment exists. Contrary to the dismal prognosis of patients with cardiac involvement of AL amyloidosis and monoclonal disorder, familial amyloidotic polyneuropathy is the result of a mutation in the gene encoding transthyretin (TTR). As of this writing, >80 amyloidogenic TTR mutations have been identified with variable cardiac involvement. TTR, a transport protein for thyroxine and retinol-binding protein, is synthesized solely in the liver. Hence, liver transplantation (with or without cardiac transplantation) can cure or at least halt disease progression. It is therefore imperative to search for hereditary forms of this disease in case biopsy-proven amyloidosis is not caused by AL or reactive systemic amyloidosis.

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To the Editor:

Singer et al report a case of biopsy-proven familial cardiac amyloidosis. In Figure 3, the authors claim that the left ventricular pressure waveform shows the dip-plateau pattern. There are several problems with this claim, as follows: (1) the fluid-filled manometry had poor fidelity with excessive oscillatory artifact; (2) the recordings included multiple ectopic beats but no steady state; (3) during long postectopic diastoles, the mid-diastolic left ventricular pressure was about 10 to 12 mm Hg, which is normal; and (4) the postectopic left ventricular end-diastolic pressure of 12 to 17 mm Hg was normal to slightly elevated, and accuracy was also limited by fidelity. Thus, although the authors nicely show cardiac amyloidosis, they do not convincingly demonstrate the full phenotypic expression of restrictive cardiomyopathy.

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