☐ DIFFERENTIAL DIAGNOSIS OF AMYLOID CARDIOMYOPATHY

☐ To the Editor:

Baca et al. urge emergency physicians to be aware of the distinction between amyloid cardiomyopathy and acute on chronic systolic heart failure, because treatment of amyloid cardiomyopathy includes loop diuretics, to the exclusion of beta-blockers, angiotensin-converting enzyme inhibitors, calcium channel blockers, and digoxin (1). I would add that amyloid cardiomyopathy should also be distinguished from constrictive pericarditis (CP) because CP requires treatment by pericardiectomy, almost to the total exclusion of diuretic therapy, especially if aggressive diuretic therapy is pursued in the absence of a suspicion for CP. The most striking example of the adverse consequence of the latter strategy is that of a 74-year-old man with unrecognized CP who became hypotensive (blood pressure 75/47 mm Hg), prone to syncopal episodes, and uremic (serum creatinine 2.1 mg/dL) as a result of aggressive diuretic therapy. The suspicion for CP was raised by the observation that his jugular venous pressure remained persistently elevated in spite of stigmata of hypovolemia. After discontinuation of diuretics and restoration of blood volume by intravenous fluids, he underwent successful pericardiectomy (2).

Although the diagnosis of CP is typically one that requires a high level of technical expertise in a sophisticated cardiothoracic center, the occasional case can be diagnosed in the emergency department on the basis of the constellation of marked elevation of the jugular venous pressure (to the angle of the jaw when the patient is sitting bolt upright), normal blood levels of natriuretic peptides, and pericardial calcification on plain chest radiography (3–6). In the context of that triad, the suspicion for CP would be reinforced by the observation that "the presence of distended neck veins in a patient who is able to lie comfortably in the recumbent posture is characteristic of the disease," especially in the presence of ascites and peripheral edema (7).

What is more challenging, however, is the ability to make a distinction between amyloid cardiomyopathy and CP. Stigmata common to both disorders were documented in a 49-year-old patient with amyloid cardiomyopathy simulating CP (8). In the latter report of amyloid cardiomyopathy simulating CP, stigmata suggestive of CP included "marked distension of the neck veins even when he was sitting up," the latter being an observation also often made in CP (3,8). In addition, "he lay flat in bed without any discomfort," the latter an observation also typical of a patient with CP (7,9). However, when the patient subsequently died, the autopsy revealed myocardial infiltration of the myocardium, and "there were no pericardial adhesions" (8).

Amyloidosis can, moreover, selectively involve the pericardium, causing CP in its own right, without involving the myocardium (9). This was the case in a 21-year-old man who presented with breathlessness and lower limb edema. On examination he had an elevated jugular venous pressure that exhibited Kussmaul's sign, the latter also a recognized feature of CP (10). Echocardiography demonstrated small ventricles with no evidence of infiltrative disease. A contrast-enhanced computed tomography scan of the chest revealed a thickened noncalcified pericardium. On magnetic resonance imaging, the myocardium did not demonstrate the characteristic contrast enhancement expected of amyloid infiltration. Cardiac catheterization showed evidence of ventricular interdependence typical of CP. Pericardiectomy was undertaken, and the histologic specimen of pericardium stained positive with Congo red, thereby validating the diagnosis of amyloid-related pericarditis (9). A patient has also been reported with amyloidosis-related CP who was shown, at autopsy, to have only minimal amyloid deposition in the myocardium and predominant amyloid deposition in the pericardium (11).

In view of the above observations, the differential diagnosis of amyloid cardiomyopathy should include the following: 1) constrictive pericarditis attributable to an etiology other than amyloidosis (3–5); 2) the association of amyloid cardiomyopathy and CP, both entities being attributable to amyloidosis (11); and 3) amyloid-related CP in the absence of concomitant amyloid cardiomyopathy (9).

Clearly, amyloidosis-related CP can coexist with amyloid cardiomyopathy whereby there is minimal amyloid deposition in the myocardium and predominant amyloid deposition in the pericardium (11). Pericardial calcification is, however, not a concomitant feature of such cases. Accordingly, even in the absence of availability of Congo red staining, the emergency physician can rule out amyliodosis if plain chest radiography or the computed tomography scan shows the presence of pericardial calcification.

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http://dx.doi.org/10.1016/j.jemermed.2019.01.040

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