Eur J Echocardiography (2002) **3,** 303–305
doi:10.1053/euje.2002.0162, available online at http://www.idealibrary.com on IDEAL®

Echocardiographic Evidence of Atrial Myopathy in Amyloidosis: A Case Report

P. S. Dardas[1]*, D. D. Tsikaderis[1], N. Mezilis[1], P. Geleris[1] and H. Boudoulas[2]

[1]*Thessaloniki Heart Institute, St Lukas Hospital, Thessaloniki, Greece;* [2]*Division of Cardiology, The Ohio State University, Columbus, Ohio, U.S.A.*

Cardiac involvement occurs in up to 50% of patients with primary amyloidosis. Diffuse amyloid deposits lead to impairment of myocardial systolic and diastolic function. Due to the severe left ventricular diastolic abnormality, left atrial contribution to left ventricular stroke volume remains critical. We report a case of primary amyloidosis where we assessed non-invasively left atrial systolic function.

(Eur J Echocardiography 2002; 3: 303–305)

Key Words: amyloidosis, left atrial function.

Introduction

Amyloid cardiomyopathy is due to deposition of amyloid in the myocardium. Cardiac involvement occurs in up to 50% of patients with primary amyloidosis[1,2]. Necropsy studies of cardiac amyloidosis typically demonstrate a thickened myocardium with enlarged chambers, predominantly the atria. Gross inspection may also reveal a rubbery consistency and the presence of intra-cardiac thrombi[3]. Diffuse amyloid deposits lead to impairment of myocardial function in two ways: replacement of functional myocardial tissue with amyloid protein that leads to reduced systolic function and diffuse infiltration of the left ventricular myocardium with inert, stiff beta-pleated protein that leads to impaired relaxation and diastolic dysfunction which usually precedes systolic dysfunction (this abnormality is sometimes called 'the stiff heart syndrome')[4,5]. Due to the severe left ventricular diastolic abnormality, left atrial contribution to left ventricular stroke volume remains critical. We report a case of primary amyloidosis where we assessed non-invasively (by using transthoracic echocardiography) multiple indices of left atrial performance.

Case Report

A female, aged 73, presented with two years' history of dyspnoea increased gradually over time. At time of presentation the patient was NYHA functional class III. Clinical examination revealed markedly elevated jugular venous pressure, right side S3 gallop, hepatomegaly, ascites and significant oedema in both lower extremities. A soft, grade I–II/VI apical systolic murmur was also heard. Electrocardiogram revealed sinus rhythm and low voltage. Biochemical profile was within normal limits. Transthoracic and transoesophageal echocardiography showed marked concentric left ventricular hypertrophy (2·0 cm) with small cavity (LVDd = 4·2 cm, LVDs = 2·8 cm) and marginally impaired left ventricular systolic function (ejection fraction = 50%) (Fig. 1(A) and (B)). Left atrium was grossly enlarged. There were left atrial spontaneous echo contrast and multiple left atrial appendage thrombi (Fig. 2). Colour Doppler showed mild mitral and tricuspid regurgitation. Transmitral Doppler flow was consistent with restrictive physiology (E wave = 91·6 cm/s, A wave = 32·8 cm/s, deceleration time = 80 ms, E/A = 3/1) (Fig. 3). Pulmonary venous flow was also consistent with restrictive physiology (systolic wave = 11·6 cm/s, diastolic wave = 39·6 cm/s, ratio <0·6)[6] (Fig. 4).

Left atrial volumes, function and work were assessed non-invasively. Left atrial volumes were calculated with biplane area–length method as determined from the two-dimensional echocardiography, using the formula $V = 8/3\pi L \times A1 \times A2$, where $\pi = 3{\cdot}14$, A1 = left atrial area in the frontal plane, A2 = left atrial area in the lateral plane and L = the shorter of the long diameters in both planes[7]. Left atrial volumes were calculated at mitral valve opening (left atrial maximal, 92 cm^3), mitral valve closure (left atrial minimum, 68 cm^3) and at onset of

**Address correspondence to:* Dr Petros S. Dardas, Vitsi 21, Panorama, Thessaloniki 55236, Greece. Tel: 0030-31-0344726; Fax: 0030-31-341828

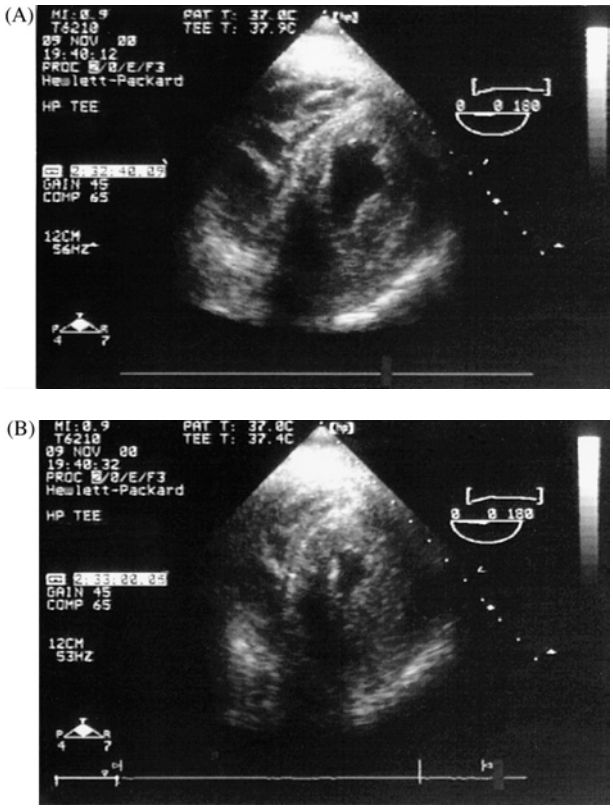

Figure 1. Transesophageal transgastric view showing marked concentric left ventricular hypertrophy with small cavity in diastole (A) and in systole (B).

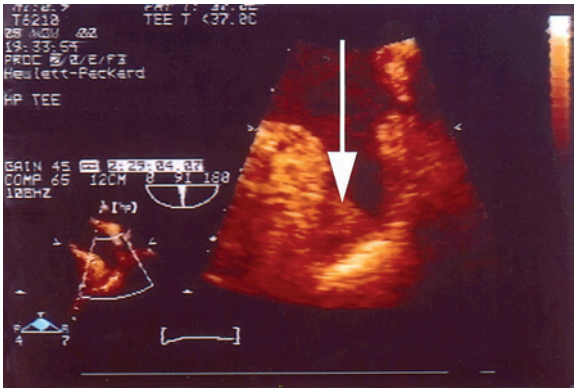

Figure 2. Transoesophageal view showing spontaneous echo contrast and multiple left atrial appendage thrombi (arrow).

atrial systole (P wave on ECG, LAP, 79 cm^3). Left atrial ejection fraction (LAEFr) was calculated by the formula LAEFr = LASV/LAP, where LASV = left atrial stroke volume = LAP − Lamin (11 cm^3). Left atrial ejection fraction was found to be 13% in our patient (mean normal values 42 ± 9, 1 SD)[7].

Left atrial kinetic energy was obtained from the formula: $\frac{1}{2} \times m \times v^2$, where m = left atrial stroke volume × ρ (ρ = blood density = 1·06 g/cm^3) and v = transmitral Doppler A velocity (32·8 cm/s)[7]. Left

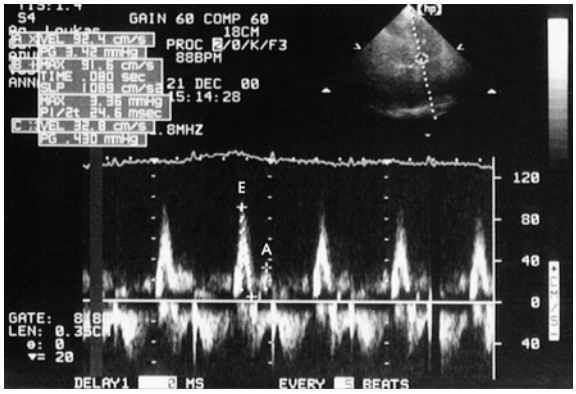

Figure 3. Transmitral Doppler flow consistent with restrictive physiology (E wave = 91·6 cm/s, A wave = 32·8 cm/s, deceleration time = 80 ms, E/A = 3/1).

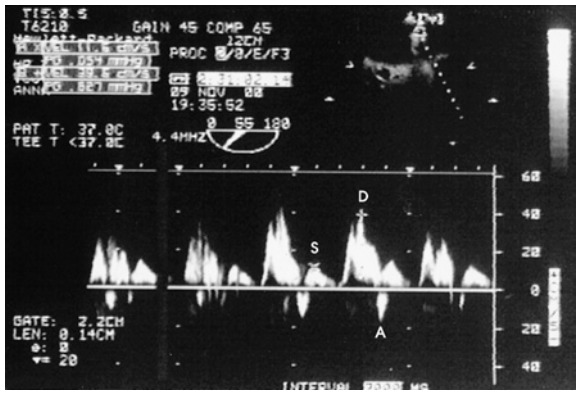

Figure 4. Pulmonary venous flow consistent with restrictive physiology (systolic wave = 11·6 cm/s, diastolic wave = 39·6 cm/s, ratio <0·6).

atrial kinetic energy was found to be 6·2 kdynes/cm (normal values for this age >25 kdynes/cm)[8].

The diagnosis of amyloidosis was confirmed by rectal biopsy. On histologic examination, the rectal biopsy specimens consisted of portions of mucosa and submucosa (Fig. 1(A)). Within the submucosa, many vessels of small and medium size were present, that contained deposits of amyloid in their walls (Fig. 5(A) and (B)).

Discussion

The presented case demonstrated left atrial myopathy in primary amyloidosis. The diagnosis of amyloidosis in this case was based on rectal biopsy and cardiac involvement was based on typical restrictive physiology as defined by transmitral and pulmonary venous flow Doppler velocities. Marked left atrial enlargement, decreased left atrial stroke volume, decreased left atrial ejection fraction and left atrial work were characteristic for this patient. Left atrial function was decreased significantly and much more than expected for the degree of left ventricular hypertrophy[9]. At the same time left ventricular systolic function was only mildly

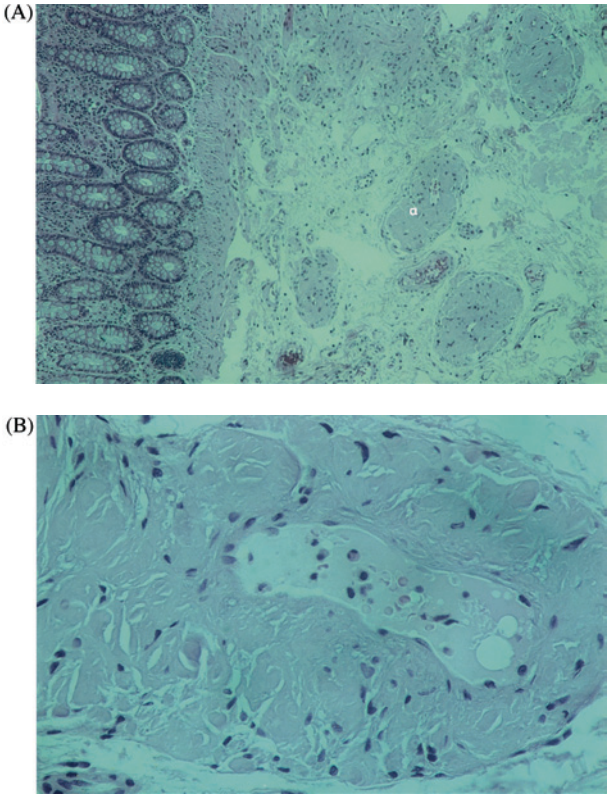

Figure 5. Low (A) and high (B) power views of rectal biopsy specimen showing amorphous eosinophilic deposits (a) in the walls of submucosa vessels, which represent amyloid.

depressed. This suggests that atrial myopathy was more profound than ventricular myopathy. Left atrial kinetic energy was used to estimate left atrial work. Previous studies have shown excellent correlation between left atrial kinetic energy and left atrial work measured directly using pressure–volume recordings[10]. Left atrial dilatation and dysfunction predisposed to intra-atrial stasis and consequently increases the risk of atrial thrombosis and thromboembolism. Furthermore, left atrial dilatation and failure with increased wall stress may result in development of atrial fibrillation. Atrial myopathy in amyloidosis may precede, be more severe than, or occur simultaneously with ventricular myopathy. Atrial failure is a major contributory factor to heart failure[11] and can possibly play a prognostic role in primary amyloidosis.

References

[1] Gertz MA, Kyle RA. Primary systemic amyloidosis — a diagnostic primer. *Mayo Clin Proc* 1989; **64:** 1505.
[2] Kyle RA. Amyloidosis. *Circulation* 1995; **91:** 1269.
[3] Roberts WC, Waller BF. Cardiac amyloidosis causing cardiac dysfunction: Analysis of 54 necropsy patients. *Am J Cardiol* 1983; **52:** 137.
[4] Swanton RH, Brooksby IAB, Davies MJ. Systolic and diastolic ventricular function in cardiac amyloidosis. *Am J Cardiol* 1977; **39:** 658.
[5] Chew C, Ziady GM, Raphael MJ, Oakley CM. The functional defect in amyloid heart disease. The 'stiff heart' syndrome. *Am J Cardiol* 1975; **36:** 43.
[6] Klein A, Cohen G, Pietrolungo J *et al.* Differentiation of constrictive pericarditis from restrictive cardiomyopathy by Doppler transesophageal echocardiographic measurements of respiratory variations in pulmonary venous flow. *J Am Coll Cardiol* 1993; **22:** 1935–1943.
[7] Boudoulas H, Boudoulas D, Sparks EA, Pearson AC, Nagaraja HN, Wooley CF. Left atrial performance indices in chronic mitral valve disease. *J Heart Valve Dis* 1995; (Suppl 2): S242–S247.
[8] Kalaras K, Sparks EA, Schuster DP, Osel K, Wooley C, Boudoulas H. Cardiovascular effects of aging. *Herz* 2001; **26:** 129–139.
[9] Triposkiadis F, Pitsavos C, Boudoulas H *et al.* Left atrial function in valvular aortic stenosis. *J Heart Valve Dis* 1992; **1:** 175–188.
[10] Stefanadis S, Dernellis J, Lambrou S, Toutouzas P. Left atrial energy in normal subjects, in patients with symptomatic mitral stenosis and in patients with advanced heart failure. *Am J Cardiol* 1998; **82:** 220–223.
[11] Hoit BD. Left atrial function in health and disease. *Eur Heart J Suppl* 2000; **2** (Suppl K): K9–K16.