

## Marked Bilateral Atrial Dilation of Unknown Cause

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### Abstract

Four elderly women with marked bilateral atrial dilation but no evidence of significant structural or functional cardiac abnormalities other than atrial fibrillation are described.

All patients demonstrated severe cardiomegaly on chest radiographs. Three patients have remained in good condition during the follow-up of 6 to 16 years except for occasional edema or pleural effusion, which was easily treated with diuretics and/or digitalis. The other patient had had only one episode of pleural effusion and leg edema at age 90 years before she died of colon cancer at age 100 years.

### Key Words

aging, atrial fibrillation, cardiomyopathies, atrimegaly, atrial dilation

### INTRODUCTION

Idiopathic right atrial dilation, which is considered to be congenital, has been described but idiopathic bilateral atrial dilation is less well reported<sup>1,2</sup>. Atrial fibrillation causes atrial dilation, but the dilation does not usually develop until gross cardiomegaly manifests on chest radiographs unless underlying cardiac disease is present. We report four patients with marked cardiomegaly associated with severe bilateral atrial enlargement but no other cardiac abnormalities except atrial fibrillation.

### PATIENTS AND METHODS

We selected four patients with gross cardiomegaly on chest radiographs but without significant structural or functional cardiac abnormalities other than bilateral atrial enlargement and atrial fibrillation.

The patients were evaluated by chest radiographs, electrocardiograms and two-dimensional echocardiograms. Left atrial volume was calculated using the ellipsoid formula by measuring the anteropos-

terior, mediolateral and superoinferior dimensions. Right atrial volume was estimated with the same ellipsoid formula, assuming equivalent anteroposterior and mediolateral dimensions<sup>3</sup>.

### RESULTS

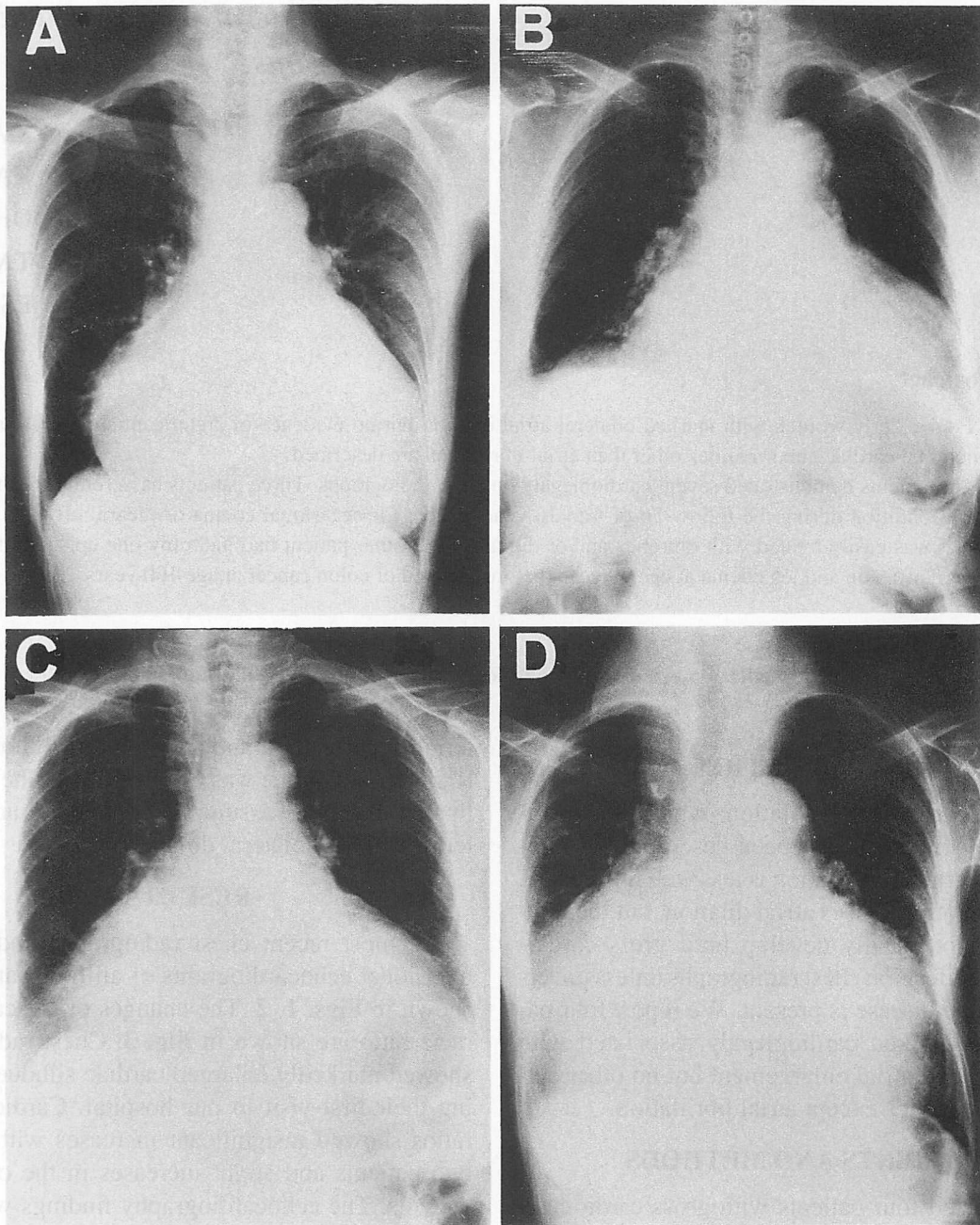
The most recent chest radiographs and two-dimensional echocardiograms of all four patients are shown in **Figs. 1, 2**. The changes of the cardiothoracic ratio are shown in **Fig. 3**. Chest radiographs showed markedly enlarged cardiac silhouettes during their first visit to our hospital. Cardiothoracic ratios showed insignificant increases with time in two patients and slight increases in the other two patients. The echocardiography findings were near normal left ventricular thickness, dimensions and ejection fraction, but a marked increase in the volume of both atrias (**Table 1**). The electrocardiograms showed atrial fibrillation in all patients since their first visit to our hospital. Right heart catheterization performed in two patients showed normal pressures. Autopsy of one patient found no evidence of a specific cardiac disease.

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Manuscript received February 17, 1995; revised March 29, 1995; accepted April 10, 1995



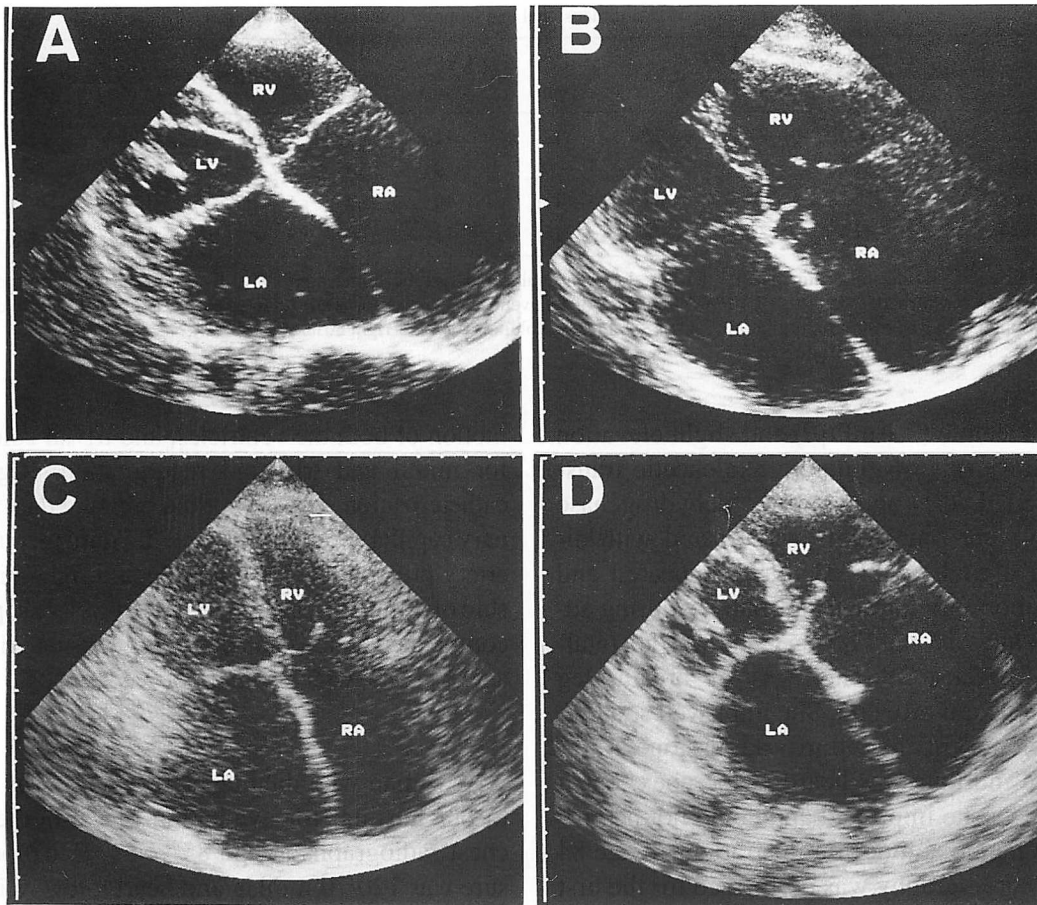
**Fig. 1** Chest radiographs

A : Case 1. B : Case 2. C : Case 3. D : Case 4.

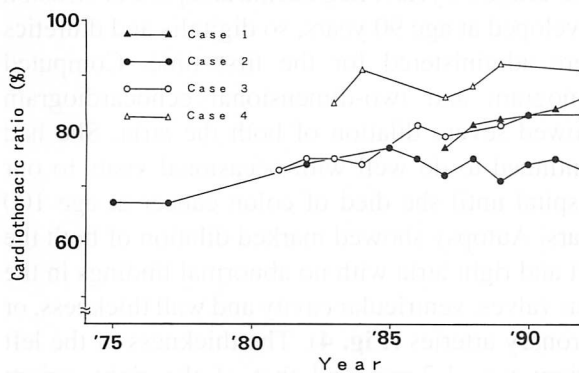
**Case 1**

An 87-year-old woman had been well until age 81 years, when she visited a physician because of shortness of breath. Cardiomegaly and pleural effusion were noted on her chest radiograph and atrial fibrillation on the electrocardiogram. Leg edema also developed later. She was referred and admitted to our hospital for further evaluation. On admission, her pulse was irregular at 70 bpm and blood pres-

sure was 129/71 mmHg. Edema was found in her legs. No rales or heart murmur were heard. Chest radiographs showed pleural effusion and marked cardiomegaly with a cardiothoracic ratio of 0.90. There were no abnormal findings in the hemogram, biochemistry or urinalysis. An electrocardiogram showed atrial fibrillation and non-specific ST-T changes in the leads II, III, aVF and V<sub>2</sub>-V<sub>6</sub>. Two-dimensional and Doppler echocardiograms re-



**Fig. 2** Four-chamber views of two-dimensional echocardiograms showing severe dilation of both atria. The scale indicates 1 cm. A: Case 1. B: Case 2. C: Case 3. D: Case 4.



**Fig. 3** Changes of the cardiothoracic ratio

vealed severe dilation of both the right and left atria but no abnormal findings in wall motion, wall thickness, size of either ventricular cavity, or in any of the four valves. Right heart catheterization performed before discharge disclosed mean pulmonary capillary wedge pressure of 6 mmHg, pulmonary artery pressure of 28/10 mmHg, and mean right

atrial pressure of 1 mmHg. The pleural effusion and leg edema disappeared and cardiothoracic ratio decreased to 0.77 soon after administration of diuretics and digitalis. A 2/6 systolic murmur, suggesting mitral regurgitation, first appeared at age 82 years. No edema or pleural effusion has not developed and no other findings of deterioration of the heart function have been observed.

### Case 2

A 92-year-old woman was diagnosed as having hypertension at age 71 years. She was referred and admitted to our hospital at age 76 years because of tachycardia with atrial fibrillation and edema in the face and lower and upper extremities. On admission, her blood pressure was 130/95 mmHg. No rales or heart murmur were audible. An electrocardiogram showed atrial fibrillation with a heart rate of 142 bpm. Chest radiographs showed pleural effusion and cardiomegaly with a cardiothoracic ratio of

**Table 1** Findings of two-dimensional and Doppler color echocardiography

	LVDd (mm)	LVDs (mm)	LVEF	IVS (mm)	LVPW (mm)	TR	MR	LAV (cm <sup>3</sup> )	RAV (cm <sup>3</sup> )
Case 1	46	27	72	12	11	+	+	106	122
Case 2	51	38	50	9	9	+	-	88	99
Case 3	38	24	68	12	10	+	+	142	203
Case 4	41	22	78	11	12	++	-	138	183

LVDd=left ventricular end-diastolic dimension; LVDs=left ventricular end-systolic dimension; LVEF=left ventricular ejection fraction; IVS=thickness of interventricular septum; LVPW=thickness of left ventricular posterior wall; TR=Doppler signal of tricuspid regurgitation; MR=Doppler signal of mitral regurgitation; LAV=left atrial volume; RAV=right atrial volume.  
- : negative, + : mild, ++ : moderate.

0.77. Hypokalemia (3.1 mEq/l) and slight elevation of the serum levels of glutamic oxaloacetic transaminase (31 IU/dl) and lactate dehydrogenase (104 IU/dl) were found. She was treated with diuretics and digitalis. The edema disappeared and abnormal laboratory findings normalized during admission. At ages 77 and 78 years, she was hospitalized again because of shortness of breath, leg edema, and pleural effusion. She improved soon after admission. On her third admission a 1/6 holosystolic murmur was heard at the apex, but it disappeared as she improved and could not be provoked with methoxamine administration. At age 84 years, echocardiography was performed for the first time and marked enlargement of both the atria was observed. She has been treated with digitalis for control of her heart rate and is in good condition, except for occasional leg edema, which has responded very well to diuretics.

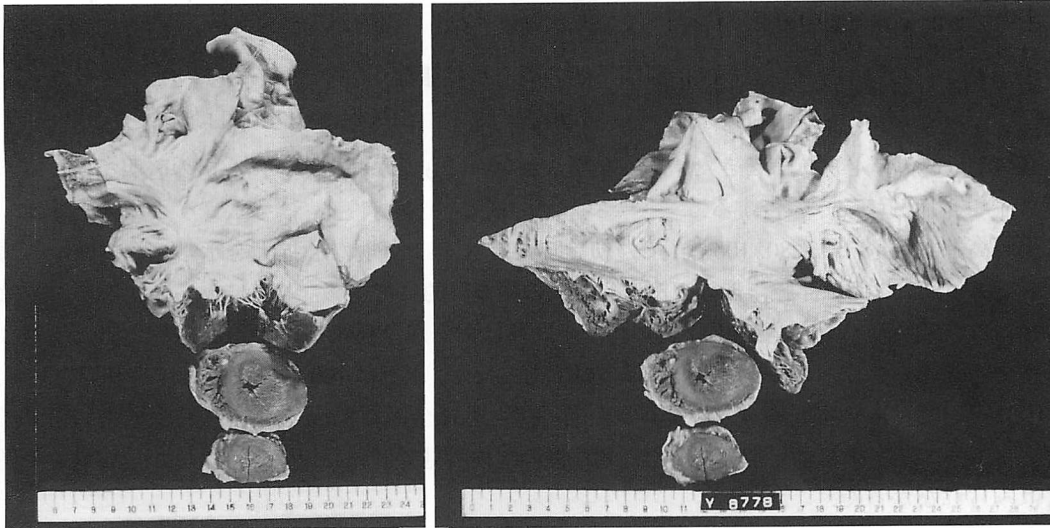
### Case 3

A currently 80-year-old woman had been hospitalized because of dyspnea at age 66 years, and was diagnosed with arrhythmia soon after. Hemiparesis of the right side and motor aphasia developed at age 69 years. Two months later she was referred to our outpatient clinic for speech therapy, when her hemiparesis had improved. At the clinic her blood pressure was 110/80 mmHg and heart rate was irregular at 90 bpm. There was no edema, and no rales or heart murmur were audible. Laboratory findings exhibited no abnormalities. Chest radiographs showed cardiomegaly with a cardiothoracic ratio of 0.73. She was admitted to our hospital for further evaluation of cardiomegaly. An electrocardiogram showed atrial fibrillation. Two-dimensional and Doppler color echocardiograms revealed bilateral atrial dilation with normal size and wall

motion of both the ventricles, and trivial color signal for mitral and tricuspid regurgitation. Right heart catheterization showed normal pressure data : pulmonary capillary wedge pressure of 7 mmHg, pulmonary artery pressure of 23/15 mmHg and right atrial pressure of 4 mmHg. She has remained well except for the complaint on admission due to digitalis intoxication at age 78 years, which soon improved.

### Case 4

A woman who died aged 100 years was referred to our hospital for evaluation of cardiomegaly on chest radiographs at age 85 years. Her blood pressure was 120/70 mmHg and heart rate was irregular at 78 bpm. There was no edema, and no rales or heart murmur were heard. An electrocardiogram showed atrial fibrillation. Cardiothoracic ratio was 0.85 at age 90 years. Leg edema and pleural effusion developed at age 90 years, so digitalis and diuretics were administered for the first time. Computed tomogram and two-dimensional echocardiogram showed severe dilation of both the atria. She had continued to do well with occasional visits to our hospital until she died of colon cancer at age 100 years. Autopsy showed marked dilation of both the left and right atria with no abnormal findings in the four valves, ventricular cavity and wall thickness, or coronary arteries (**Fig. 4**). The thickness of the left atrium was 1.2 mm and that of the right atrium 0.7 mm. Microscopic findings of the atrial walls showed marked irregular fibrosis and degeneration of myocardial fibers with granular cytoplasm and large nuclei. Vacuolation was occasionally observed in the myocardial fibers. A decreased number of myocardial fibers were recognizable in the atrial walls. Congo red staining gave a negative reaction in the atrial walls. Microscopic findings of the ventricular walls showed no significant abnor-



**Fig. 4** Macroscopic specimen of Case 4 showing marked dilation of both right and left atria compared to the ventricular cavities

*Left* : view of the left atrium.

*Right* : view of the right atrium.

malities.

## DISCUSSION

Atrial fibrillation is associated with atrial enlargement<sup>3</sup> although whether atrial fibrillation is the cause or consequence is still debated<sup>4</sup>. Our four cases may suggest a disease which affects mainly the atria, causing atrial dilation and atrial fibrillation, or may be merely extreme cases of atrial dilation caused by atrial fibrillation.

Previous reports have discussed the relationship between atrial volume, duration of atrial fibrillation, and age. Sugiura *et al*<sup>5</sup> reported no correlation between the duration of atrial fibrillation and atrial volume from the evaluation of 169 autopsy cases with atrial fibrillation. Garber *et al*<sup>6</sup> found the amplitude of f waves decreased as age increased, but there was no correlation between the amplitude of f waves and left atrial dimension, indirectly showing an insignificant correlation between age and atrial size. Petersen *et al*<sup>7</sup> measured the anteroposterior dimension of the left atrium of the patients with atrial fibrillation of short duration (within 3 months) and of long duration (1 year or more), finding a significantly larger dimension in patients with atrial fibrillation of long duration,  $43 \pm 5$  and  $49 \pm 5$  mm,

respectively. Echocardiography repeated 6 months later showed an increase in the dimensions of both groups, to  $49 \pm 6$  and  $53 \pm 5$  mm, respectively. However, when corrected for body surface area, there was no significant difference between the two groups.

These reports suggest the atrium increases in size rather rapidly at the beginning of atrial fibrillation, and thereafter the rate of dilation decreases to a limiting value of atrial dilation. In our four patients it was uncertain when atrial dilation began. No case has atrial fibrillation advanced to atrial standstill, which is also associated with cardiomegaly and atrial dilation<sup>8</sup>. The autopsy findings of Case 4 showed fibrosis and degeneration of myocardial fibers of the atrial walls, but no evidence of valvular heart diseases, ventricular or coronary artery disease. The other three patients were followed up at our outpatient clinic for 6 to 16 years, and have not shown significant deterioration except for occasional leg edema or pleural effusion, which respond to therapy very well. Our cases may suggest the presence of a disease, which mainly affects the atria and causes atrial enlargement like dilated cardiomyopathy, which mainly affects the ventricles.

## 要 約

## 原因不明の両心房拡張症

前田 茂 加藤 洋一 大川真一郎 田久保海誉 江崎 行芳

心房細動以外、心房拡張をきたすような異常がないにもかかわらず、両心房の著明な拡張のみが目立つ4症例につき報告する。

全例高齢女性(87, 92, 80, 100歳)で、初診時よりX線上著明な心拡大を示し、心電図は心房細動であった。2例は浮腫、他の2例は心拡大の精査目的にて受診。初診時からの4例の経過は6, 16, 12, 16年で、3例は経過中浮腫と胸水の既往はあるが、利尿剤によく反応し、他の1例は心不全の既往もなく、4例ともに経過は良好であったが、1例は100歳で大腸癌により死亡した。心エコー図では、房室弁輪よりの奥行は左右心房ともに全例で8cmを越え、左室拡張終期径、左室駆出率、左室壁厚は正常であった。死亡例の剖検所見では、心房壁が薄いほか、特異的な異常はみられなかった。

心房細動では一般に心房は拡大するが、経年的に増大を続けるという報告はなく、これらの症例は心房細動による心房拡大の極端な例をみているのか、あるいは心房に特異的に影響を及ぼす病態を考えるべきなのか、今後検討されるべき症例と思われる。

*J Cardiol* 1995; 26: 33-38

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