Extreme Bilateral Atriomegaly

Review of the Literature and Report of a Case

By Wayne R. Rogers, M.D. and Benjamin Wittels, M.D.

Extreme enlargement of the atria is unusual and its mechanism is not clearly established. A case is reported here of extreme bilateral atriomegaly with some special features of interest and the entire problem is reviewed.

THE cardiac atria of the normal adult are nearly equal in size; the right averages 163 ml. in capacity, the left 140 ml.\(^1\) The left atrium is more prone to enlarge, usually in association with mitral valve disease; it rarely reaches extreme volumes of 3,000 ml.\(^2\) Extreme left atriomegaly was first reported as “aneurysmal dilatation of the left auricle” in 1849 by Hewett,\(^3\) but the clinical aspects have been appreciated less than 50 years. Excellent reports have been made by Bramwell and Duguid\(^4\) and by Daley and Franks.\(^5\) However, little attention has been given to the status of the right atrium in these patients, since it was usually only slightly dilated.

Observation of a patient with a left atrium 2,000-ml. size and a right atrium of 800-ml. size stimulated a review of the literature aimed at evaluating the clinical and pathologic aspects of bilateral extreme atriomegaly. Cases were arbitrarily included for study only if there was anatomic evidence that the volume of the left atrium exceeded 1,000 ml. and that of the right atrium, 500 ml. Nine such cases were found, and their relevant data are listed in table 1.

Case Report

The patient (B.I.H. A55-170) was a white man, 35 years old at death. At 4 years of age recurrent pains and swelling developed in the extremity joints. At age 9 these symptoms caused his hospitalization at the House of the Good Samaritan, Boston, for 13 months. Examination disclosed chorea, cardiac enlargement, a loud mitral presystolic murmur, softer mitral and aortic systolic murmurs, and a very soft aortic diastolic murmur. Electrocardiograms showed normal sinus rhythm, P-R intervals of 0.19 to 0.24 sec., notching of P\(_2\), and an electric axis of +75°. Improvement occurred on rest and diet therapy. Final diagnoses were active rheumatic carditis, mitral insufficiency and stenosis, and possibly aortic insufficiency.

The first recorded cardiac fluoroscopy, done at age 12, disclosed “well marked enlargement” of the left atrium, which formed the upper right border of the enlarged, mitral-shaped, cardiac silhouette. At age 18, atrial fibrillation developed abruptly with a ventricular rate of 130/min. Subjectively only palpitation was noted and this subsided following digitalization. On 0.1 Gm. of the whole leaf daily, he maintained a ventricular rate of 80 to 90/min. thereafter. At age 19, cardiac fluoroscopy disclosed enlargement of the right atrium and right ventricle. The cardiothoracic ratio was 14.5:24.2 cm. His general condition remained good as confirmed by semi-annual medical examinations. He was able to carry out a life of moderate activity, working full time as a draftsman.

In January 1950, at age 29, he entered the Beth Israel Hospital, Boston, complaining of the gradual development of exertional dyspnea, orthopnea, paroxysmal nocturnal dyspnea, occasional precordial aching unrelated to exertion, nonproductive coughing, and rare episodes of dysphagia for a period of 4 months. Examination disclosed a tall, thin male of medium build. The rectal temperature was 99.6 F.; respirations were labored at a rate of 30/min.; the apical pulse was irregular at a rate of 108/min.; and the arm blood pressure was 160/100 mm. Hg. (All subsequent blood pressure determinations were within normal limits.) The point of maximum cardiac impulse was located in the sixth intercostal space in the left anterior axillary line. There were loud, rough apical systolic and diastolic murmurs and thrills, softer aortic systolic and diastolic murmurs, a loud pulmonic second sound, and a soft aortic second sound. There was slight cyanosis but no clubbing of the fingers. The neck veins were not distented; the lungs were clear; the liver edge was palpable; and edema was absent. Laboratory studies including complete blood counts, plasma nonprotein nitrogen, Hinton serologic reaction, and urinalysis were normal except for 2+ to 4+ albuminuria. The arm-to-tongue circulation time was 32 sec. The vital capacity was 2.1 L. (normal 4.3 L.). Chest fluoroscopy disclosed enlargement of all 4 cardiac chambers,
most markedly of the left atrium, which pulsed synchronously with ventricular systole. No intracardiac calcification was seen. The esophagus was displaced posteriorly and to the left, and the bronchial angle at the carina was widened by the enlarged left atrium. There was dilatation of the pulmonary artery, the lungs appeared slightly congested, and the costophrenic angles contained a small amount of fluid. Electrocardiograms showed atrial fibrillation, right axis deviation (+120°), and probable right ventricular hypertrophy. Treatment with rest, low-salt diet, continuation of digitalis, and mercurial diuretic injections every other day resulted in lessening of dyspnea and a 12-pound weight loss, whereupon he was discharged.

Because of gradual relapse on this regimen he was rehospitalized from March to May 1950. Additional studies disclosed an antecubital venous pressure of 185 mm. of saline and an icterus index of 28 units. Basal metabolic rates averaged +5 per cent, and the 24-hour thyroid uptake of radioiodine131 was 37 per cent. As a final therapeutic measure, this euthyroid patient was given 32 mc. of radioiodine in 2 divided doses in April 1950.

He changed little until September 1950, when myxedema appeared. Thereafter, exertional capacity improved considerably, and mercurial injections were required only every 2 weeks. Chest roentgenograms during this period showed slight diminution in cardiac size and clearing of pulmonary congestion (fig. 1). In January 1952, thyroid, 6 to 18 mg. daily, was instituted to alleviate symptoms of myxedema. The basal metabolic rate was maintained at an average of −25 per cent and the serum cholesterol level at 280 mg. per cent. He was able to return to full-time work for the next 4 years.

In August 1955 exertional dyspnea reappeared. In October the appearance of mild coughing and of chills and fever necessitated rehospitalization. The cardiac findings were unchanged. There were now neck vein distention, dulness, decreased breath sounds and crepitations over the right lower posterior chest, moderate hepatomegaly, and slight leg edema. No evidence of thrombophlebitis was found. The chest roentgenogram showed a more greatly enlarged heart and consolidation of the right middle lobe that was interpreted as a possible pulmonary infarct. The electrocardiogram disclosed increased evidence of right ventricular hypertrophy (fig. 2).

In conjunction with the cardiac therapy, heparin was administered intravenously every 4 hours in 50 mg. doses. Dyspnea increased, cyanosis, hypotension, stupor, and periodic breathing developed, and he died on the third hospital day.

Postmortem examination revealed 200 ml. of serous fluid in the pericardium and dense fibrous adhesions between the left atrium and the pericardial sac. The heart was huge because of enormous enlargement of the atria and auricular appendages (fig. 3). The left atrium was uniformly dilated and contained more than 2 L. of blood. It abutted against the lateral wall of the left hemithorax and extended to within 5 cm. of the right chest wall. Superiorly, it reached to the level of the aortic arch;

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Table 1.—Clinical and Pathologic Features of Ten Cases of Extreme Bilateral Atrio-megaly

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at death (yr.)</th>
<th>Heart wt. (Gm.)</th>
<th>Atrial vol. (ml.)</th>
<th>Valve Lesions</th>
<th>Valve orifice</th>
<th>Fibrosis</th>
<th>Infarct</th>
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<td>2000</td>
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<td>1300</td>
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<td>25</td>
<td>17</td>
<td>&lt;17</td>
<td>&lt;28</td>
<td>760</td>
<td>1235</td>
<td>550</td>
</tr>
</tbody>
</table>

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* LA = left atrium.
† Estimations are based on linear measurements.
‡ Capsules denote pronounced valvular lesions; MS = mitral stenosis; MI = mitral insufficiency; ti = tricuspid insufficiency; t. inc. = tricuspid incompetency; as = aortic scarring.
§ memb. = membranous.
Fig. 1. Posteroanterior and lateral chest roentgenograms in 1954. LA is left atrial border. RA is right atrial border. Note the marked retrodisplacement of the barium-filled esophagus and the absence of pulmonary congestion.

Fig. 2. Electrocardiogram of October 25, 1955. There is a deep S wave in leads V3 and V6, that is not clearly visible.

inferiorly, it rested upon the diaphragm; and posteriorly, it widened the main stem bronchial angle and displaced the esophagus to the left and posteriorly. It compressed and markedly displaced both lungs posterolaterally. The fossa ovalis was closed, and it was enlarged 3 to 4 times normal. The endocardium displayed diffuse reticulation and was heavily encrusted with calcium plaques. They were especially prominent in and about the fossa ovalis and the auricular appendage. The walls, generally translucent, were thin, fibrous, and seemingly depleted of muscle. The right atrium was about one third the volume and one-half the width of the left. Its endocardium was focally thickened but devoid of calcification. The pectinate muscles appeared hypertrophied. There was marked hypertrophy and dilatation of the ventricles. The left ventricular wall had a maximum thickness of 1.7 cm., the right 0.6 cm., and the interventricular septum, 2.2 cm. There was no mural thrombus. The
mitral valve was stenotic, rigid, and heavily calcified. The cross-sectional diameters of its orifice were 1.5 by 0.7 cm. There were no vegetations. The chordae tendineae were thickened, fused, and shortened. The tricuspid valve showed slight thickening and rolling of the free borders, and a few of its chordae tendineae were slightly thickened and shortened. The aortic valve was normal except for minimal commissural fusion between 2 of its cusps. The pulmonic valve was unremarkable. The circumference of the mitral valve ring was 14.5 cm., of the tricuspid 12 cm., aortic 9 cm., and of the pulmonic 8 cm. The coronary arteries, examined by the injection and dissection technic of Schlesinger, had a few discrete, nonstenosing atheromatous plaques in the main left and right stems. The arterial supply of the atria appeared commensurate with the atrial size. Prior to fixation the total weight of the empty heart was 688 Gm., the free left ventricle weighed 150 Gm., free right ventricle 120 Gm., interventricular septum 125 Gm., left atrium 118 Gm., right atrium 54 Gm., interatrial septum 18 Gm., and epicardial fat 63 Gm.* The heart had not deformed the chest markedly.

Histologically, the atria showed widespread fibrosis. In both ventricles there were several Aschoff bodies and numbers of small arteries showed slight degrees of medial hypertrophy with metallocysis. There was slight fibrosis of the myocardium, involving mainly the inner layers of the left ventricle.

The right lung weighed 980 Gm. and the left 480 Gm. The right middle and lower lobes were largely consolidated by pneumonitis. There was very striking pulmonary atherosclerosis, especially in the lobar arteries. The lobar arteries of the right middle and left lower lobe each contained an organizing thrombus 2.5 to 3.0 cm. long; in the latter location about 75 per cent stenosis was produced. No fresh thrombi were found. There was bilateral hydrothorax (right 350 ml., left 50 ml.) and ascites of 350 ml. The liver weighed 1360 Gm. and was very firm and finely nodular. Both kidneys were scarred. The thyroid gland consisted of 2 Gm. of fibrous tissue. No phlebothrombosis was found in the pelvic or leg veins.

Final pathologic diagnoses were rheumatic heart disease, moderately active, (a) mitral stenosis and insufficiency, (b) commissural fusion of the aortic valve, mild, (c) cardiomegaly, 688 Gm., (d) enormous atria with fibrosis and calcification; chronic passive congestion of the lungs; chronic recurrent pneumonitis of the right middle and lower lobes; pulmonary atherosclerosis, severe; pulmonary emboli; cardiac cirrhosis of the liver; healed renal infarcts; thyroid atrophy following radioiodine therapy.

**Discussion**

**Terminology.** The structure involved in the enlargement is the entire atrium, including the appendage. The processes involved are mainly dilatation with varying degrees of muscular hypertrophy or atrophy and fibrosis. The dilatation is relatively uniform; however, owing to uneven resistance presented by adjacent structures, the extremely dilated atrium may assume an asymmetric contour. This finding has led to the use of the description "aneurysmal dilatation." The term "massive dilatation" is somewhat inappropriate since the atrial wall itself is usually not much increased in weight; "enormous" or "extreme dilatation" is preferable. Usually this degree of enlargement of the left atrium is best judged clinically

* Mean weights of normal adult male hearts, divided according to this technic, are listed by the pathologist, Leo Reiner, M.D., as follows: left ventricle 120 Gm., right ventricle 80 Gm., both atria 80 Gm., interventricular septum 90 Gm., total heart 320 Gm.
by finding its right border extending beyond the structures that normally form the right cardiac silhouette in the posteroanterior projection of the roentgenogram.

Pathologic Aspects. Eight patients (cases 1–8) had chronic rheumatic heart disease with severe mitral valve involvement. A rather uniform picture of confluent scarring of these leaflets and chordae tendineae was found converting the valve into a tough fibrous septum; gross calcification was noted in 4 of these valves. The oval or round ostia would admit 1 fingertip to 3 fingers, and they were evidently both insufficient and stenotic. Combined ventricular hypertrophy of moderate to marked degree was present in all 8 cases. In none was functionally significant aortic or pulmonic valvular disease or myocardial disease thought to be present, although in 4 the aortic valve was scarred.

In case 9 the solitary cardiac lesion noted was pure mitral insufficiency due to a rent 18 mm. long in the anterior leaflet resulting from the impact of a pistol slug. Over a 10-year period, it produced enormous dilatation with hypertrophy of all 4 cardiac chambers.

Case 10 was quite exceptional in that there was no significant organic valvular lesion. The atrial dilatation appeared to be due solely to chronic constrictive pericarditis, which acted presumably by producing prolonged atrial hypertension.

The pathogenesis of enormous left atrial dilatation is not well understood. Nearly always a marked and long-standing rheumatic mitral valve lesion appears to be the major etiologic factor, although structurally nothing unique has been described about the valve lesions that have been found associated with the largest atria. Occasionally there is tight stenosis, but more commonly a considerable element of insufficiency is evident. Recently, left atrial catheterization in 15 patients with "giant" left atria has disclosed a single feature that correlates with the enormity of the atrial enlargement. This feature consists of "rapid and severe vibratory fluctuations of pressure" coincident with a regurgitant jet, which suggested the possibility of a poststenotic mechanism in reverse, as a factor in the production of the enormous dilatation.

The right atrial dilatation in these patients can be attributed in large part to the effect of rheumatic tricuspid insufficiency in 4 and to tricuspid incompetence, due to stretching of the tricuspid ring in chronic right heart failure, in the remaining 6 cases. The right atrial size was less than half that of the left atrium in each instance except case 2, in which the right atrium was almost as large as the left. This patient was the only one who had a moderate degree of tricuspid stenosis in conjunction with insufficiency. A similar stenotic and insufficient tricuspid valve was present in association with the largest right atrium on record (2,150 ml). Curiously, the latter patient's left atrium was normal in size despite the presence of definite mitral stenosis. In the present rheumatic patients, the relatively smaller size of the right atrium as compared to the left is consonant with the lesser severity of the tricuspid than the mitral lesion, with the more severe fibrosis of the left atrium, and with the fact that, in most, the disease of the left side of the heart can be assumed to be of longer duration.

Atrial dilatation may be facilitated by primary disease of its wall. Evidence of atrial disease other than dilatation was lacking in 6 patients; therefore, if there is a local etiologic factor augmenting dilatation, it appears not to be essential. Possible stigmata of left atrial rheumatic infection were described in the form of extensive fibrosis in 4 patients, in 2 of whom there was marked endocardial calcification; the right atrium of 2 showed moderate fibrosis. In 5 patients, pericardial adhesions were noted on the left atrium, although in none was there evidence of traction that caused aneurysm. That these adhesions were rheumatic in origin, and were not due alone to trauma involving the surface of the enlarging atrium, is suggested by their absence in case 9 in which there was no evidence of rheumatism. Atrial Aschoff bodies were not found in any case. Comparison of these atrial findings in the 8 rheumatic patients with those usually found post mortem in chronic rheumatic valvular disease is only somewhat suggestive of unusually severe arthritis in 4. A history of recurrent or severe manifestations of acute rheumatic fever was noted in 5. Nonrheumatic atrial disease, particularly septal defect, was not implicated in
any case. The coronary arteries were described in 4 hearts, and all were normal. Injection studies of the atrial branches of case 1 disclosed a structurally adequate arterial system.

The relationship of chronic atrial fibrillation to the atriomegaly in these patients seemed to be more one of result than cause. Fibrillation was probably absent in the patient with the largest left atrium, and in case 1, the left atrium was known to be enormously enlarged 7 years before fibrillation developed.

Clinical Aspects. Symptoms and signs arose primarily as a result of congestive heart failure or atrial pressure on adjacent structures. Therefore, in the early stages discomfort was minimal. In case 1 when the left atrium had dilated to form the upper right border of the cardiac silhouette, the patient continued for 17 years to have mild exertional dyspnea as the only symptom. This phenomenon of minimal symptoms despite marked cardiomegaly is both characteristic and unique in this type of heart disease. It has been attributed to a reservoir function of the large left atrium, which has been demonstrated anatomically and fluoroscopically to be distensible. Probably of greater importance is good ventricular functional capacity, which is in keeping with the good condition of the ventricular myocardium and vasculature found post mortem.

Congestive heart failure gradually developed in all patients. Its course was lengthy, ranging from 3 to 18 years and averaging 8.8 years. Initially the evidence of failure was left-sided (dyspnea) in 7, right-sided (dependent edema) in 1, bilateral in 1, and unknown in 1. Eventually it was definitely bilateral in 9 and probably so in the tenth case. Evidence of cardiac cirrhosis was described in all except cases 2 and 4. Satisfactory response to routine therapy including digitalis was noted in 6. Radioiodine-induced hypometabolism effected excellent relief of the medically intractable heart failure in case 1 for a period of 4 years.

Symptoms related to left atrial enlargement, which in part overlap those of heart failure, were noted in 5 patients. For instance, the enlarged heart may fill the lower half or more of the thoracic cavity; hence respiratory embarrassment may occur from reduction of vital capacity, a feature that has not been studied.

Cough associated with left or right bronchial displacement or compression was noted by 4 patients; and in case 1, bronchial compression appeared to be responsible for the chronic pneumonitis. Hemoptysis was recorded in 1 case, possibly resulting from pulmonary infarction. Mild dysphagia, presumably from esophageal compression, was noted by 2 patients. Precordial pain irregularly related to exertion was described in 2 patients, neither of whom had occlusive coronary artery disease at postmortem examination. Injection of the coronary arteries disclosed well vascularized atria and ventricles in case 1; however, there was good clinical and pathologic evidence of pulmonary arterial hypertension to which the pain may have been related. That the mechanism of production of such pain may involve atrial ischemia was suggested in a patient with an enormous left atrium and fibrillation whose pain was relieved by rest or nitrroglycerin; yet it was not associated with electrocardiographic changes, which suggest that the pain was not of ventricular origin. In 1 patient hoarseness was attributed to the left atrial dilatation. Recorded effects of left atrial pressure that were not noted in these patients are: (1) superior vena caval obstruction, (2) collapse of the left lung due to main stem bronchial compression on the aorta, and (3) erosion of thoracic vertebral bodies. The irregular occurrence of pressure effects from these extremely large atria suggests that their development is not particularly dependent on the degree of atriomegaly. No specific pressure effect was ascribed to the dilated right atrium.

The occurrence of a mural thrombus in an enormously dilated atrium is not unexpected in view of the common presence of rheumatic endocardial damage and of stasis. Such a thrombus was found post mortem in cases 2 and 8, and each was located in the right atrium and was small in size. Infarcts possibly due to embolization from the heart were found in systemic organs in 4 patients, and pulmonary infarcts were found in 3.

Electrocardiograms made during advanced stages of the heart disease were described in 4 patients. All were abnormal tracings and in all there was atrial fibrillation. Two showed the pattern of right ventricular hypertrophy. In
the remaining 2, ventricular hypertrophy was not evident; in 1 there was right axis deviation, in the other the electric axis was normal.

All patients died in congestive heart failure. Death was precipitated by pneumonitis in 1, by pulmonary embolism in 1. Two died suddenly, presumably of cardiac arrhythmia, since autopsy revealed no adequate anatomic cause. Subacute bacterial endocarditis was not encountered.

The age at death of the present rheumatic patients ranged from 28 to 48 years, averaging 37. Similar average longevity (34 years) and similar average duration of heart failure (7.5 years) occurred in Smith and Levine’s group of 32 patients with tricuspid and mitral rheumatic disease who were selected without regard to atrial size. Thus no life-prolonging protective function can be ascribed to the enormous atria of the present patients. In contrast, these authors found that mitral disease alone (stenosis) permitted a longer life, and the associated heart failure averaged only 2.9 years in duration with medical treatment.

These considerations of extreme atrial dilatation are heightened in importance by the recent performance of mitral valvuloplasty in several patients having “giant” left atrioventricular dilatation, the concomitant presence of right atrial dilatation is of small additional clinical significance, usually indicating a more advanced state of heart disease with tricuspid valvular incompetency.

**Acknowledgment**

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**Summary**

The clinical and pathologic aspects of bilateral enormous atrial dilatation are presented in 1 case and reviewed in 9 cases in the literature.

The left atrial dilatation was related to rheumatic mitral insufficiency and stenosis in 8 cases, to traumatic pure mitral insufficiency in 1 case, and to chronic constrictive pericarditis in 1 case. The less marked right atrial dilatation was related to rheumatic tricuspid insufficiency in 4 cases and to tricuspid incompetence due to chronic right ventricular failure in 6 cases. Little evidence was found to implicate other pathogenic factors such as atrial wall disease; the mechanism by which some atria dilate so greatly remains obscure.

In persons having enormous left atrial dilatation, the concomitant presence of right atrial dilatation is of small additional clinical significance, usually indicating a more advanced state of heart disease with tricuspid valvular incompetency.

**References**


Thromboembolic phenomena may represent a serious complication of the use of corticotropin and cortisone. Such complications were observed in 5 patients with hypercholesteremia, idiopathic or secondary to other disease, suggesting a causal relationship between the pre-existing elevated serum cholesterol and the thromboembolic phenomena. Only 1 of these patients had clinically evident pre-existing cardiovascular disease. It is suggested that the hypercoagulability induced by the corticosteroids superimposed on the vascular disease that may be associated with this metabolic error, predisposes the individual to thromboembolic phenomena. Caution in the administration of steroid therapy to hypercholesteremic individuals is advised. The results in 1 of these cases suggests that prophylactic anticoagulant therapy may be helpful.

Kitchell
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