Giant Left Atrium in Rheumatic Heart Disease: A Report of 18 Cases Treated by Mitral Valve Replacement

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EXTREME enlargement of the left atrium, usually referred to as giant, gigantic or aneurysmal dilatation, has long been known and reported in the literature to be an uncommon if not a rare finding associated with rheumatic valvular disease. The reported incidence of giant left atrium (G.L.A.) is about 0.3% in those suffering from rheumatic heart disease.[3] Although there are no accepted criteria, most authors define cases according to x-ray appearance whereby the left atrium forms the right border of the heart shadow and approximates the right chest wall.[3, 11, 18] An alternative definition is where both the left and right borders of the heart shadow are formed by the left atrium.[19] We restricted our cases to those fulfilling either of these criteria, confirmed by actual operative findings.

Early reports of G.L.A. appeared as autopsy findings [4, 8, 14, 15] and the associated mitral lesion was noted. Later, the association with rheumatic heart disease was established [5] and a fairly typical clinical picture was described with a high incidence of mitral insufficiency.[1, 2, 16] With the advent of cardiac catheterization more accurate information was possible,volume measurements and hemodynamic studies being performed.[10, 13, 17, 20] No significant correlation could be found between the size of the left atrium and other measurements, such as left atrial pressure, calculated diameter of the mitral valve opening, pulmonary arterial or capillary pressure [13] and the amount of mitral regurgitation.[7] Cardiac surgery gave a new impetus to the study of these cases since most patients have advanced cardiac disease and incapacity, and are candidates for mitral valve surgery.

Material

In the 3-year period 1967–69, eighteen patients who underwent open heart mitral valve surgery were found to have GLA. The total number of patients operated upon for rheumatic mitral valve disease in this period was 150 (120 "open-heart" procedures and 30 "closed" commissurotomies). Thus in this series the incidence of GLA is 12% of all patients operated upon for mitral valve disease. Table 1 shows the pertinent findings in these patients. Representative x-ray studies are illustrated in Figs. 1, 2, 3, and 4. All were in advanced chronic heart failure with grade IV incapacity (N. Y. Heart Association). There were 12 females and 6 males ranging in age from 6–62 (average age 30). All the patients had mitral insufficiency. In nine (50%) it was the predominant lesion, whereas in the others, mitral stenosis was more obvious. Pure mitral stenosis, however, was not found; but seven can be said to have had pure mitral insufficiency. Either associated tricuspid insufficiency or stenosis or both

* Deceased.

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TABLE 1. *Pertinent Findings in 18 Patients*

No.	Sex	Comm.*	Age	Diagnosis**	Date of Op.	Operation	Outcome/Condition
1	F	Druze	12	MI, TI	Jan. 1967	M valve repl. disc T annuloplasty	Late death cerebral embolus
2	M	Arab	6	MI, MS	17.1.68	M valve repl. disc	Hospital death ventric. fibrillation
3	F	Indian	16	MI	24.1.68	M valve repl. disc	Late death embolus cerebral and coronary
4	M	Iraqi	50	MI	6.3.68	M valve repl. disc	Improved
5	M	Polish	57	MS, MI, TI	8.4.68	M + T valve repl. double disc	Much Improved
6	M	Polish	36	MS, MI	1.7.68	M valve repl. Disc	Improved
7	M	Roumanian	30	MS, MI	8.7.68	Triple valve M. T. A. repl. 2 discs + 1 ball	Unimproved
8	F	Arab	27	MI, TI	29.7.68	M valve repl. disc T annuloplasty	Well
9	M	Arab	36	MS, MI, TS, TI, AI mild	23.12.68	M + T valve repl. disc	Hemodynamically improved severe anxiety state
10	F	Arab	17	MS, MI	21.4.69	M valve repl. heterograft	Very well
11	F	Yugoslav	62	MS, MI	7.5.69 4.6.70	M valve repl. heterograft M valve repl. disc	Late syst. murmur insuff. valve
12	F	Arab	26	MS, MI	14.5.69	M valve repl. heterograft	Hospital death ventric. fibrillation
13	F	Arab	19	MI	16.6.69	M valve repl. heterograft	Very well
14	F	Israel	36	MI, MS	6.8.69	M valve repl. disc	Improved
15	F	Arab	24	MI, TI	13.8.69	M valve repl. disc	Improved
16	F	Arab	27	MS, MI, TI	27.8.69	M valve repl. heterog. T annuloplasty	Improved
17	F	Israel	34	MS, MI, TS, Ti, AI mild	29.9.69	M + T valve repl. heterograft	Hospital death low output synd.
18	F	Arab	22	MI, TI mild	27.10.69	M valve repl. heterograft	Very well

* Community origin.
** Predominant lesion first.

was found in seven patients; aortic insufficiency only in one. Fourteen patients had single mitral valve replacements performed (nine Kay Shiley disc valves, five aortic heterografts). In three of these tricuspid annuloplasty was added. Three double valve replacements, tricuspid and mitral, were undertaken (two with disc valves and one with aortic heterografts). One triple valve replacement, aortic, mitral and tricuspid is also included in this series.

Operative Technic

The heart was approached via mid-line sternotomy incision. Extracorporeal circulation was maintained by the Rygg-Kyvsgaard* disposable bubble oxygenator. Body

* Polystan, Copenhagen.

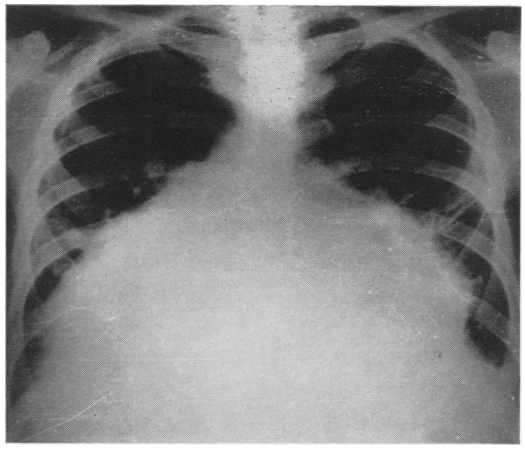

FIG. 1. Patient 5: Massive dilatation of L. A. is seen in all directions in a 57-year-old man.

temperature was cooled to 30° C. The mitral valve was replaced prior to other valvular surgery. When isolated mitral valve surgery was necessary, the approach was directly into the left atrium via the inter-atrial groove. In those requiring mitral and tricuspid valve surgery, the mitral valve was approached through the right atrium and inter-atrial septum. No procedures to reduce the volume of the left atrium were performed.

Results

There were three hospital deaths (16.6% mortality) and two late deaths from systemic emboli giving a total cummulative mortality of 27.7%. Thirteen survivors have been under observation for up to 3½ years. Twelve are clinically and hemodynamically improved, many strikingly. One, patient 7, following triple valve replacement, remains in chronic congestive heart failure. Patient 11 underwent disc replacement of a malfunctioning heterograft valve 13 months after the first operation. Figs. 5, 6, 7, show some of the postoperative results.

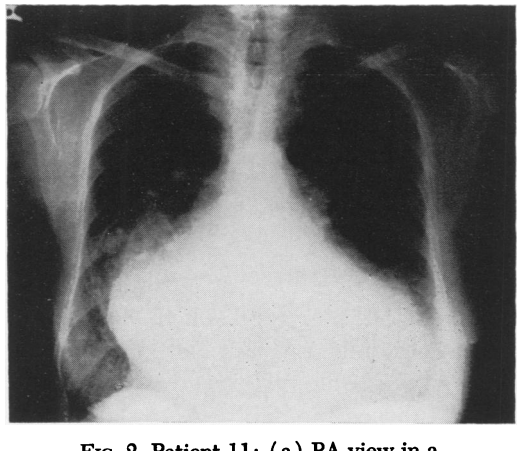

FIG. 2. Patient 11: (a) PA view in a 62-year-old woman.

Discussion

The number of reported series of surgically treated cases of GLA is small:

Author	Year	No. of Cases	Procedure
Venner[21]	1954	1	Mitral valvotomy
Kent[10]	1956	5	Mitral valvotomy
De Sanctis[3]	1964	2	1 Mitral valvotomy 1 Mitral valve replacement
Guest[6]	1965	1	Mitral valve replacement
Johnson[9]	1967	17	16 mitral valve replacement + plication. 1 plication L.A. only
Piwnica[18]	1967	29	Mitral valve replacement
Knudsen[11]	1969	3	Mitral valve replacement
Present report	1970	18	Mitral valve replacement
Total		76	

In our series GLA was found in 12% of all cases operated upon for mitral valve disease, including open and closed procedures. This high percentage may be due to the fact that a large proportion of our patients undergo cardiac surgery in advanced stages of disease, and no patient with rheumatic valvular disease has been denied surgical therapy on the grounds that the cardiac lesion is too far advanced. Three children (aged 14, 12 and 6) with GLA, reflect the early onset and severity of rheumatic disease in our region. All adults had long rheumatic and cardiac histories. Severe cardiac cachexia was a striking clinical feature, a fact also stressed by Piwnica.[18] Table 2 shows the height and weight of our patients. This contradicts the

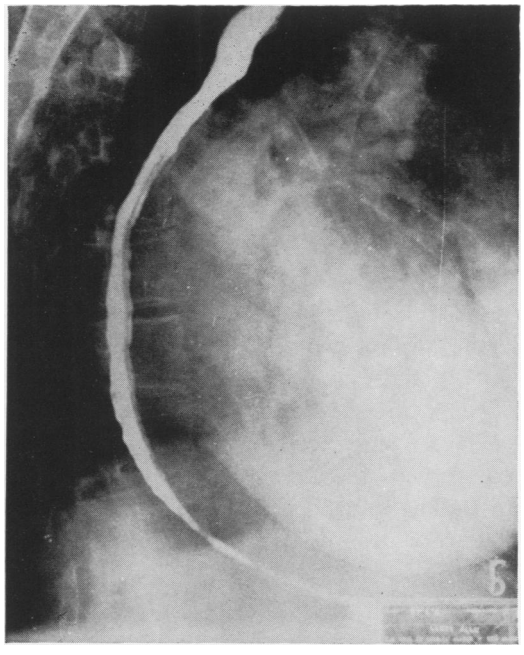

FIG. 2. (b) Lateral view shows, in particular, the marked posterior displacement of the esophagus.

report of De Sanctis [3] who considers patients with GLA to have a relatively benign course even when not operated upon.

Seven patients had right heart catheterizations performed preoperatively, and, in all, intra-operative pre-bypass pressure measurements were taken, except when we were forced to hurriedly connect to the heart-lung machine because of deteriorated cardiac function. The results (Table 3) show that the left atrial pressure was nearly always markedly raised. Pulmonary arterial pressures were in general elevated, in most above 50 mm. Hg systolic. Johnson [9] found only one patient with resting pulmonary aterial pressure above 50 mm. Hg in a series of 17 cases. No obvious correlation was found between the size of the atrium and the intra-atrial pressure.

No patient had clinical or laboratory evidence of rheumatic activity. No Aschoff bodies were found in biopses of the atrial appendages. Marked fibrosis and chronic inflammation were the main histologic

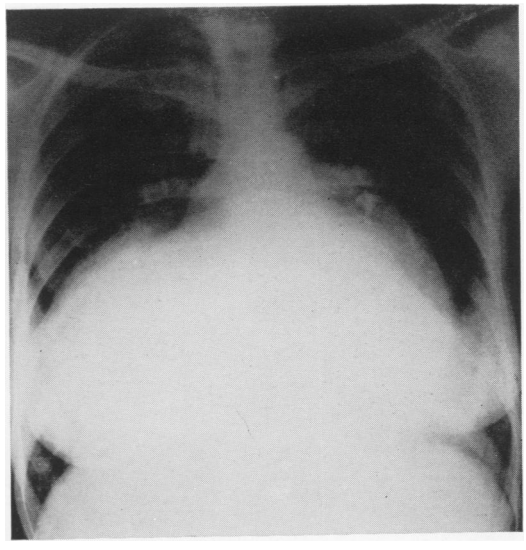

FIG. 3. Patient 14: Massive enlargement of the L. A. has almost reached both chest walls in a 36-year-old woman.

findings which may explain the high incidence of atrial fibrillation—all adults exhibiting this rhythm. Yet no clots were found in the left atrial cavities of any patient. It is possible that the turbulence, caused by the mitral regurgitation, is the

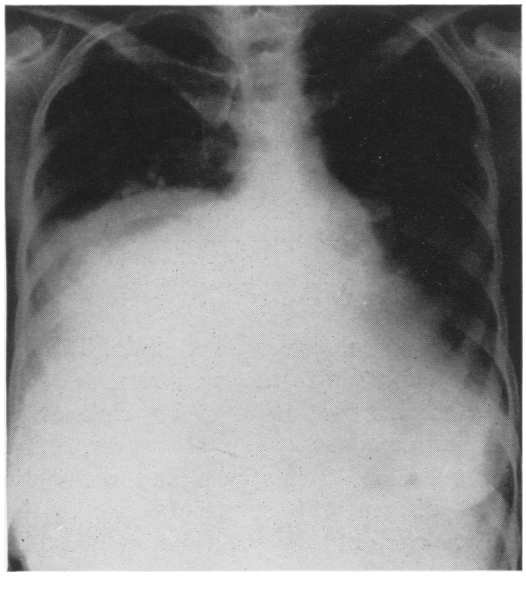

FIG. 4. Patient 17: Typical aneurysmal dilatation of the L. A. is seen particularly to the right in a 34-year-old woman.

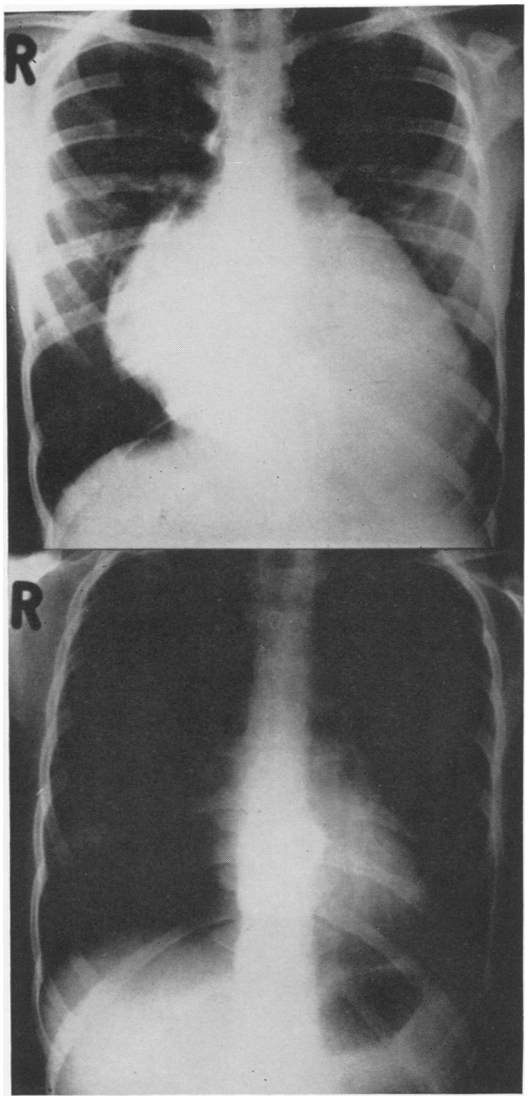

FIG. 5. (a & b). Patient 3: Pre- (top) and post-operative (bottom) chest x-rays of a 14-year-old girl. This patient died 15 months after operation due to embolic complications from the disc valve.

reason for the absence of thrombi. The extreme dilatation of the left atrium is probably related to the jet of insufficiency, a finding common to all. However, other factors, such as long standing rheumatic heart disease with severe damage to the myocardium of the left atrium, may also be responsible for the GLA.

The most striking operative finding was the vast balloon-like enlargement of the left atrium which occupied the whole of the posterior, right and left mediastinum. The rest of the heart sat on this balloon and was displaced forward like a "crown on a cushion." The left atrial wall was a thinned out fibrotic-like structure. No attempt was made to diminish the size of the left atrium by plication or excision as advocated by Johnson[9] or Le Roux.[12] There does not seem to be sufficient evidence that this procedure is of benefit. In our opinion, prolongation of the cardiopulmonary bypass time in these patients does not warrant such a step except when the atrium so compresses the lung as to produce atelectasis. The mitral valve was always replaced with no attempted repair as most of the valves were so damaged by the rheumatic process that repair was not feasible. In addition, the precarious hemodynamic status of these patients required a procedure which would guarantee a perfect hemodynamic result. Nevertheless, discontinuation of extracorporeal circulation was difficult

TABLE 2. *Height and Weight of 18 Patients with G.L.A.*

Case No.	Height Cm.	Weight Kg.	Estimated Weight* Kg.
1	130	26.50	28
2	105	15	17.50
3	164	42	50
4	159	64	65
5	161	63	67
6	174	52	73
7	180	67	79
8	160	43	55
9	168	59	64
10	152	39	47
11	146	48	57
12	157	45	54
13	162	54	54
14	153	40	54
15	152	44	49
16	157	40	54
17	160	42	58
18	180	55	66

* According to Height-age tables; build and blood pressure studies Vol. I, Chicago 1959 (Scientific Tables Geigy Basle 1962).

in many instances and isoproterenol or adrenalin infusion was frequently necessary in the postoperative period. It is our impression that these patients with GLA came to operation with considerable myocardial damage. Confirmatory evidence for this is

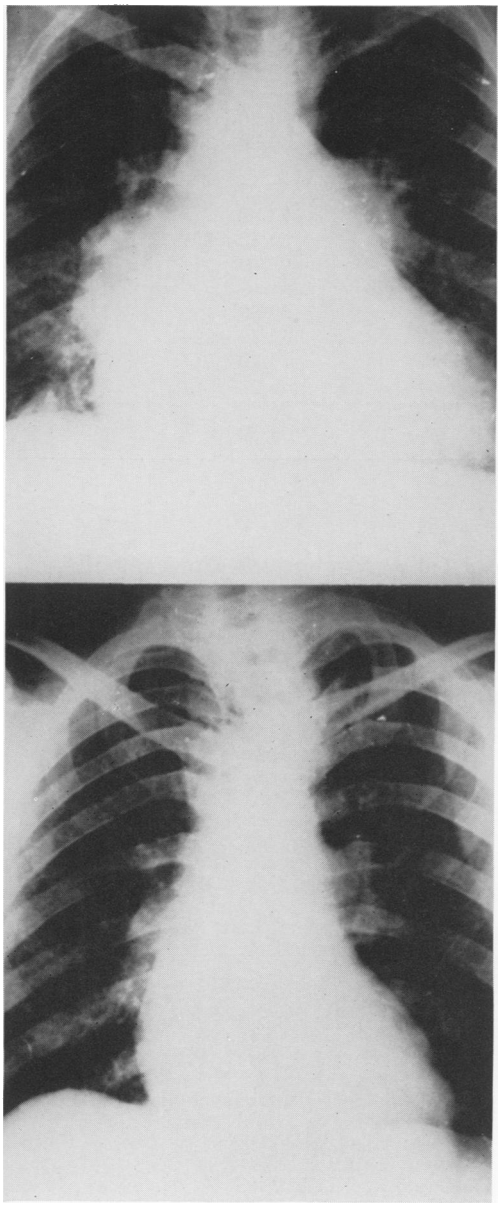

FIG. 6. (a & b). Patient 9: Pre- and postoperative chest films show a decrease in size of the L. A. after mitral and tricuspid replacement in a 36-year-old man.

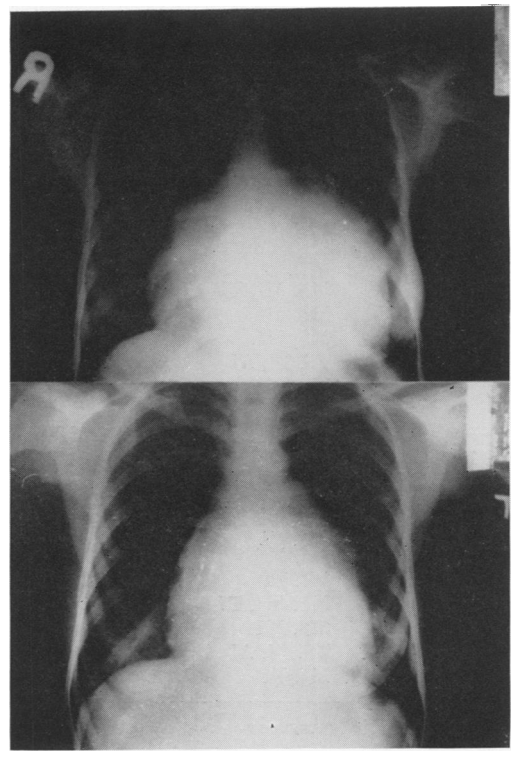

FIG. 7. (a & b). Patient 15: Globular enlargement of the L. A. and postoperative results are seen in a 24-year-old woman.

the relatively long period elapsing before the full benefit of the operation is apparent. Patients above 30 years of age particularly, required supportive digitalis and diuretic treatment for many weeks, even months, after the operation. The marked cachexia, however, responded more rapidly to surgical therapy with return of appetite and weight gain soon after departure from the hospital. The long-term results in these extremely ill patients give cause for satisfaction. Twelve patients (67% of the series) are considered as having good to excellent results, most having returned to their former employment. One patient following triple valve replacement has not responded well to operation and is still in congestive cardiac failure. As there is no evidence of valve malfunction, his condition is probably of myocardial or endocardial origin.

TABLE 3. *Left Atrial, Pulmonary Artery, Right Atrial and Systemic Pressure Measurements in the 18 Cases of G.L.A.*

No.	Diagnosis	Pressure mm. Hg			
		Left Atrium	Right Atrium	Pulmonary Artery	Systemic
1	MI, TI	60/5-m25		50/30	
2	MI, MS		a 10 m 7*	64/30-m35*	128/85*
3	MI	70/20-m25	13/8	70/30	90/70*
4	MI	35/10-m20	10/5	35/20	
5	MS, MI, TI		19/4-m14*	55/26-m33*	100/60*
6	MS, MI	70/45-m60	10/2-m5	90/50-m70	
7	MS, MI, TS, TI, AI	50/20-m30		50/20-m35	116/77*
8	MI, TI	40/15-m28	20/0-m12	40/20-m34	
9	MS, MI] TS, TI, AI	35/5-m22		55/25	75/?
10	MS, MI	70/20-m45		70/40	110/65
11	MS, MI			60/30-m35	
12	MS,MI	38/20	18*	95/60*	97/75*
13	MI	20/0-m10	35/11-m20		100/60
14	MI, MS	55/25-m38	14	50/30	120/70
15	MI, TI	22/10	15/5	30/10-m22	80/0
16	MS, MI, TI	40/25	19*	75/40	80/*
17	MS, MI TS, TI, AI	26/20-m25	19/10-m14	56/22*	80/55
18	MI, TI	75/35-m45	12/5-m10	90/50	110/80

* Preoperative measurements at catheterization.

Summary

Eighteen patients with giant left atrium were operated upon. These cases form 12% of 150 consecutive mitral valve operations. Mitral insufficiency due to rheumatic heart disease was the dominant lesion in all. All patients were in advanced chronic congestive heart failure and cardiac cachexia. Atrial fibrillation was constantly present except for three children. Left atrial and pulmonary artery pressures were raised in all patients, very high levels being recorded in most.

Mitral valve replacement was carried out in all patients. In addition, six patients had tricuspid replacement or repair and one required replacement of the tricuspid and aortic valves. Procedures to reduce the size of the left atrium were not employed.

Fifteen patients survived operation. Two of these died later as a result of thromboembolic phenomena related to the prosthetic valve. Twelve of the 13 long-term survivors are strikingly improved. Consequently, cardiac surgey should be recommended in instances of GLA and severe mitral valve dysfunction notwithstanding the advanced, often preterminal condition of the patients.

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