The Heart Surgery Forum #1997-8127
1 (1):71–75, 1998

Online address: www.hsforum.com/vol1/issue1/1997-8127.html

Autotransplantation Procedure for Giant Left Atrium Repair

(#1997–8127 ... December 23, 1997)

Ugolino Livi, MD[1] and Giulio Rizzoli, MD[2]

[1]Division of Cardiothoracic Surgery and Transplantation Unit, University of Padova, Italy
[2]Division of Cardiothoracic Surgery, University of Padova, Italy

ABSTRACT

Background: Giant left atrium has been associated with bronchopulmonary and left ventricular compression [Kawazoe 1983].

Case Report: We present a patient with severe congestive heart failure (CHF), respiratory insufficiency and a giant left atrium (GLA) following two previous mitral valve procedures and tricuspid valve annuloplasty in the distant past. Mitral prosthetic function and ventricular systolic function were felt to be normal leading to a tentative diagnosis of diastolic restriction from left ventricular compression and pericardial constriction. A pericardial decortication procedure through left thoracotomy was initially done but proved ineffective. Subsequently, full evidence of hemodynamic failure due to the giant left atrium and its respiratory complication was recognized and the patient underwent cardiac autotransplantation procedure [Kosak 1987], with the aim to reduce the left atrial dimensions to normal.

Conclusions: Calcification of posterior left atrial wall prevented a completely satisfactory reduction of atrial size and the severity of ventricular adhesions from the previous pericardial procedure resulted in very long cardiopulmonary bypass time with severe bleeding complications. This case provides ample evidence that GLA can cause respiratory failure and needs to be surgically corrected.

INTRODUCTION

Giant Left Atrium (GLA) is a well-known, albeit rare, complication of mitral valve disease [Root 1990, Badui 1995]. Patients with mitral valve disease and extreme enlargement of the left atrium usually exhibit significant decrease in chamber size following corrective mitral valve surgery. Occasionally progression of atrial distention has been observed following mitral valve replacement even in the absence of prosthetic valve malfunction or tricuspid valve disease [Mahapatra 1983].

Giant left atrium can be responsible for respiratory dysfunction by compressing the left main bronchus or the right middle and lower lobes or by causing hemodynamic disturbances due to distortion of the atrioventricular connection and compression of the posterobasal left ventricular wall.

We present the case of a patient with huge atrial enlargement and congestive heart failure (CHF) despite a normally functioning mitral prosthesis with a discussion of the surgical options for correction.

CASE REPORT

A 47 year old male was referred to our hospital with severe relapsing CHF. Pertinent history included a mitral valve annuloplasty using a #32 Carpentier ring plus a tricuspid valve annuloplasty of the posterior leaflet with tricuspid bicuspidalization in 1975 at a Paris hospital. The operation was performed with hypothermic ventricular fibrillation at 28 degrees centigrade without aortic cross clamping. The operative record described the left atrium as "very enlarged". After the operation, mean left atrial pressures (LAP) averaged 15 mmHg. There was a 1/6 systolic thrill evident. Follow-up echocardiographic evaluation demonstrated some residual mitral incompetence and stenosis.

In 1986, the patient underwent mitral valve replacement with a 29 mm Medtronic Hall prosthesis along with a DeVega tricuspid annuloplasty at a nearby center. This procedure was performed with moderate hypothermia, aortic clamping and crystalloid cardioplegia. The mitral valve was both stenotic and incompetent due to chordal fusion and papillary shortening. The left atrium was noted to be huge, with displacement of the heart entirely into the left chest cavity. The immediate postoperative course was stormy with low output syndrome requiring high inotropic dosage. He recovered and was clinically well until 1995 although follow-up echo showed persistent pulmonary hypertension with estimated PA pressures of about 45 mmHg.

Address correspondence and reprint requests to Giulio Rizzoli, MD, Cardiochirurgia, Via Giustiniani 2, 35128 Padova, Italy

© 1998 Forum Multimedia Publishing, LLC

Upon presentation to our institution, physical examination showed massive hepatomegaly (5 cm below the transverse umbilical line) without any obstruction of inferior vena caval orifice [Minagoe 1992] and severe bilateral leg edema. Pulmonary testing revealed normal diaphragmatic movement but severely restricted ventilation from external compression with a functional capacity of 41% of predicted and effort limitation due to severe dyspnea. Chest x-ray revealed a massive cardiac silhouette (See Figure 1, ◉).

Computerized Axial Tomography (CAT) scan showed a giant left atrium which was partially calcified at the bottom and contained a stratified posterior thrombus. The ventricles lied on top of it, pressed to the thoracic wall (See Figure 2, ◉).

Transesophageal echo showed normal prosthetic function, some left ventricular dilatation (102 ml EDV, 42 ml ESV) with normal systolic function and moderate ventricular dysfunction due to diastolic restraint. Right ventricular function was normal but there was 3+ to 4+ tricuspid incompetence. The left atrium was gigantic with a volume estimated between 2 and 3 liters with spontaneous left atrial echocardiographic contrast [Voci 1991] and an extensive organized posterolateral thrombus.

Cardiac catheterization revealed pulmonary hypertension (PAp 81/22/42), a cardiac index of 4.4 L/min/m2 and a left ventricular ejection fraction of 52%. Total pulmonary arteriolar resistance was 5 Woods units. The left ventricular silhouette was pressed against the lateral thoracic wall, so that the left ventricular axis was completely vertical and the mitral prosthetic ring is positioned at 90 degrees with respect to it (see Figure 3, ◉).

Ten days after admission, the patient underwent a left lateral thoracotomy to release suspected left ventricular pericardial adhesions and to mobilize the left ventricle from the lateral thoracic wall. In spite of the absence of pericardial thickening, the pericardium was removed as far as possible with the hope of relieving any possible cardiac compression. Pulmonary artery pressures at the end of operation were 35/17 mmHg, left atrial pressure was 16 mmHg and right atrial pressure 15 mmHg.

On postoperative day 4 he developed oliguria, impressive CHF and required reintubation along with ultrafiltration. Bronchoscopy revealed subtotal stenosis of the left main bronchus and atelectasis of the left lung. The possibility of bronchial stenting was evaluated and excluded by the thoracic surgeon in the face of persisting left atrial compression. On postoperative day 15 a tracheostomy was placed. He remained in respiratory and renal failure for another 43 days, but eventually came off dialysis and partially off ventilatory support. At that time (58 days following his pericardiectomy), thorough discussions regarding definitive correction of his left atrial enlargement were undertaken with the patient and a surgical decision reached to reduce left atrial size by cardiac autotransplantation along with surgical correction of the tricuspid insufficiency.

The operation began with institution of cardiopulmonary bypass via left femoral artery and venous cannulation. The chest was opened through a median sternotomy after unloading the right atrium and ventricles which were adherent and compressed to the sternal plate. The entire ventricular mass was severely adherent to the atelectatic left lung. It was very difficult to find a dissection plane as a consequence of the relatively fresh and edematous adhesions secondary to prior pericardial decortication operation 58 days earlier. Finally the aorta was clamped and asystolic arrest induced with 1,500 milliliters of antegrade cold blood cardioplegia (4 degrees Celsius). The aortic and pulmonary artery were divided and the right and left atrium divided as in a heart transplant cardiectomy (see Figure 4, ◉).

The whole heart was removed and placed in an iced basin (see Figure 5, ◉). The mitral prosthesis was fully mobile without any pannus ingrowth and the previous de Vega annuloplasty was reasonably competent.

A large amount of the lateral left atrial wall was removed but we were forced to leave behind a large amount of the posterior wall which was severely calcified and adherent to the posterior mediastinum (see Figure 6, ◉).

Finally the heart was reimplanted using a continuous 4–0 Prolene® to suture the heart to a narrow rim of uncalcified atrial wall. Significant left atrial reduction to an estimated volume of 500–600 ml was obtained.

The aorta was crossclamped for a total of 125 minutes and the heart function reassumed after a warm cardioplegic shot and a single cardioversion. The total elapsed bypass time was 8 hours and 20 minutes. After 20 minutes partial bypass was resumed for further 85 minutes because of major bleeding despite recovery of ventricular function. Fifteen hours after start of the procedure the patient was returned to the ICU hemodynamically unstable and with persistent severe bleeding. He expired several hours later from low output syndrome.

DISCUSSION

The hemodynamic relevance of atrial size reduction is not clear. In fact after reduction the atrium will remain an immobile reservoir in most cases unless restoration of sinus rhythm can be achieved. Analogously in the presented case we were initially not able to define the role of atrial enlargement in determining or contributing to this patient's CHF. Given an apparently normal mitral prosthetic function and echocardiographic evidence of preserved left ventricular systolic function with some diastolic restraining, we hypothesized that pericardial release could increase the compliance of the left ventricle and eliminate a possible cause of limited left ventricular filling and cardiac output [Yamamoto 1992]. In the absence of discrete constrictive pericarditis, this maneuver did not improve cardiac performance but instead aggravated respiratory dysfunction by causing total left lung atelectasis and worsening of the patient's CHF symptoms

The role of atrial reduction in the setting of mitral valve replacement has been widely discussed in the literature both in regard to its ventilatory effect as well as its hemodynamic effects but a consensus has not yet been reached.

Much of the present knowledge is due to Kohei Kawazoe [Kawazoe 1983] who in the early 80s outlined three major effects of massive atrial enlargement:

1. Paradoxical movement of the posterobasal wall of the left ventricle, compressed and bent inward by the downward extension of the left atrium.
2. Filling of the left ventricle during diastole in an unphysiologic direction. Both these factors are responsible for hemodynamic failure and of an increased incidence for low output syndrome in the postoperative mitral patient.
3. Respiratory disturbance yielded by the compression of left main bronchus from upward extension by the left atrium or compression of the right middle lobe by rightward extension of the left atrium.

A further hemodynamic effect due to obstruction of the inferior caval outflow has also been described [Minagoe 1992]. Therefore Kawazoe [Kawazoe 1983] advocated the use of a paraannular and superior plication technique to reduce left atrial size in all patients presenting to mitral valve repair or replacement and GLA. This suggestion has been subsequently criticized by authors who failed to demonstrate an independent effect of GLA on the requirements of postoperative respiratory care [Matsuda 1990] or on the risk of mitral valve surgery [Di Eusanio 1988].

On the contrary Piccoli [Piccoli 1984] and Otaki [Otaki 1993, Otaki 1994] showed a statistically significant incremental risk effect of GLA in their series. In discussing a case presentation, Hara [Hara 1993] also expressed the opinion that plication is very beneficial in patients with GLA who have signs of compression of the bronchus and lungs. The benefits of this surgical procedure have been quantified in terms of left atrial dimensions and respiratory function by Hagihara [Hagihara 1995] and Isomura [Isomura 1993]. Finally in this setting atrioplasty has been liberally applied by Russian [Konstantinov 1990] and Ukrainian surgeons as well [Matchk 1997].

Differences in judgment among surgeons is probably related to the fact that the entity GLA has never been quantified. We have no doubt that the definition fits our case in whom an estimated atrial volume of at least 2000 ml was found. GLA is defined as atria above 300 ml of volume or 60 mm diameter [Matsuda 1990] and this could explain the different findings concerning hemodynamic derangement and incremental risk.

In our patient, congestive heart failure and respiratory failure occurred with normal left and right ventricular function and a normally functioning mitral prosthesis. Therefore we were forced to explain it as a consequence of the left atrial enlargement and in concert with Kawazoe's postulated pathophysiology. Having reached this uneasy conclusion, the next step was to define the proper treatment modality. Atrial reduction would have been the patients third sternotomy and the risk of gaining exposure to the distended atria and ventricles was rather high. Our surgical group was divided between those favoring a limited operation involving an intraatrial plication of Kawazoe's type through the enlarged left atrial roof and those advocating an autotransplantation procedure to more effectively reduce the size of the left atrium.

In our opinion, Kawazoe's operation had the primary advantage of a limited dissection although serious doubts were raised about the feasibility of effectively reducing a lateral atrial wall reaching all the way to the left thoracic cage. We were also concerned that there were no available data on the success of this approach in patients undergoing a 3rd reoperation and with dense adhesions of surrounding structures to the atrial wall. The second choice (which was theoretically appealing) required the isolation of the entire heart, a risky procedure which is nonetheless routinely performed in our unit where we have a large experience with reoperations and heart transplantation. Autotransplantation has received intensive investigation in the animal laboratory. It was successfully applied to the treatment of otherwise unresectable human cardiac tumors since 1987 [Kosak 1987, Scheld 1988]. More recently autotransplantation has been applied to the treatment of the long QT syndrome [Pfeiffer 1992]. Kitamura [Kitamura 1993] has proposed this technique for repair of complex cardiac anomalies. It appeared to be a natural option in this patient with essentially normal ventricles sitting on top of an enormously dilated left atrium. Transplantation often requires tailoring of the recipients left atrium to adapt to donor heart size.

What was not properly taken into account was the proximity (58 days) of the previous pericardial decortication operation. Fresh edematous adhesions to the left lung resulted in a very long total elapsed bypass time and severe bleeding complications. It was also not fully appreciated that the calcified plate, which was clearly visible above the spine on the CT scan, would have prevented resection and mobilization around the pulmonary venous orifices, limiting the desired size reduction.

In retrospect, we were faced with a patient condemned by a natural history that could not possibly be interrupted by a less invasive operation. Autotransplantation procedure was the sole chance of success if only adhesion severity would have allowed a reasonable and usual operative time. This case and its tragic conclusion makes also a case for prevention of this severe complication during conventional mitral valve surgery by primary plication of enlarged left atria, even at the expense of an additional 20 to 30 minutes of aortic crossclamping time [Shigenobu 1992, Kawazoe 1983].

REFERENCES

1. Badui E, Delgado C, Enciso R, Graef A, Solorio S, Madrid R, Cruz H. Silent giant left atrium. A case report. Angiology. 46(5): 445–8, 1995.
2. Di Eusanio G, Gregorini R, Mazzola A, Clementi G, Procaccini B, Cavarra F, Taraschi F, Esposito G, Di Nardo W, Di Luzio V. Giant left atrium and mitral valve replacement: risk factor analysis. Eur J Cardiothorac Surg. 2(3): 151–9, 1988.
3. Hagihara H, Kitamura S, Kawachi K, Morita R, Taniguchi S,

© 1998 Forum Multimedia Publishing

Fukutomi M, Kawata T, Hasegawa J, Yoshida Y. Left atrial plication combined with mitral valve surgery in patients with a giant left atrium. Surg Today. 25(4): 388–42, 1995.
4. Hara Y, Honda T, Nakamura H, Hamasaki T, Kobayashi T, Ishiguro S, Sasaki S, Kuroda H, Mori T. A case of left atrial plication for mitral valvular disease with giant left atrium Kokyu To Junkan 41(2): 193–6, 1993.
5. Isomura T, Hisatomi K, Hirano A, Maruyama H, Kosuga K, Ohishi K. Left atrial plication and mitral valve replacement for giant left atrium accompanying mitral lesion. J Card Surg. 8(3): 365–70, 1993.
6. Matchk Zenyk and Ivaniv Yuri. Personal communication, Aug. 1997.
7. Kawazoe K, Beppu S, Takahara Y, Nakajima N, Tanaka K, Ichihashi K, Fujita T, Manabe H. Surgical treatment of giant left atrium combined with mitral valvular disease. Plication procedure for reduction of compression to the left ventricle, bronchus, and pulmonary parenchyma. J Thorac Cardiovasc Surg. 85(6): 885–92, 1983.
8. Kitamura N, Yamaguchi A, Miki T, Noji S, Otaki M, Kurata A. Autotransplantation as optimal technique for recurrent malignant myxoma of left ventricle. Nippon-Kyobu-Geka-Gakkai-Zasshi. 41(3): 445–51, 1993.
9. Konstantinov BA, Cherepenin LP, Tarichko IuV, Rasulov IR, Nechaenko MA, Bobkov VV, Shevelev II. Atrioplasty in surgical correction of mitral valve defect complicated by left-sided atriomegaly Grud Serdechnososudistaia Khir. (2): 3–8 , 1990.
10. Kosak M, Gabrijelcic T, Brecelj A, Erzen V, Kocbek B, Obrez I, Medvescek N, Bardofer I, Rutar V, Ovcak Z. The world's first successful autotransplantation of the heart for recurrent tumor. Acta-Chir-Iugosl. 34(2): 131–9, 1987.
11. Mahapatra S, Goldberg MJ, Ryan JM, Rubenfire M, Cascade PN. Progressive extreme biatrial enlargement following mitral valve replacement. Chest. 84(3): 306–8, 1983.
12. Minagoe S, Yoshikawa J, Yoshida K, Akasaka T, Shakudo M, Maeda K, Tei C. Obstruction of inferior vena caval orifice by giant left atrium in patients with mitral stenosis. A Doppler echocardiographic study from the right parasternal approach Circulation. 86(1): 214–25, 1992.
13. Otaki M, Kawashima M, Yamaguchi A, Tamura H, Kitamura N. Surgical treatment of cardiac cachexia with mitral valve disease: the effect of preoperative IVH and left atrial plication on postoperative respiratory condition Kyobu Geka. 46(2): 117–20, 1993.
14. Otaki M. Surgical treatment of patients with cardiac cachexia. An analysis of factors affecting operative mortality. Chest. 105(5): 1347–51, 1994.
15. Pfeiffer D, Fiehring H, Warnke H, Pech HJ, Jenssen S. Treatment of tachyarrhythmias in a patient with the long QT syndrome by autotransplantation of the heart and sinus node-triggered atrial pacing. J Thorac Cardiovasc Surg. 104(2): 491–4,1992.
16. Piccoli GP, Massini C, Di Eusanio G, Ballerini L, Iacobone G, Soro A, Palminiello A. Giant left atrium and mitral valve disease: early and late results of surgical treatment in 40 cases. J Cardiovasc Surg Torino. 25(4): 328–36, 1984.
17. Root JD, Hammerman AM, Fischer KC. The CT appearance of the giant left atrium. Clin Imaging. 14(4): 305–8, 1990.
18. Scheld HH, Nestle HW, Kling D, Stertmann WA, Langebartels H, Hehrlein FW. Resection of a heart tumor using autotransplantation. Thorac Cardiovasc Surg. 36(1): 40–3, 1988.
19. Shigenobu M, Takagaki M, Kohmoto T, Okada T, Senoo Y, Komoto Y, Teramoto S. Effect of left atrial plication for the giant left atrium on left ventricular function. Acta Med Okayama. Jun, 46(3): 189–93, 1992.
20. Voci P, Scibilia G, Bilotta F, Maugeri B, Caretta Q, Mercanti C, Marino B, Reale A. Spontaneous left atrial echocardiographic contrast in mitral stenosis: early disappearance after valve replacement. J Am Soc Echocardiogr. 4(6): 648–50 , 1991.
21. Yamamoto K, Masuyama T, Tanouchi J, Uematsu M,Doi Y, Naito J, Hori M, Tada M, Kamada T. Decreased and abnormal left ventricular filling in acute heart failure: role of pericardial constraint and its mechanism. J Am Soc Echocardiogr. 5(5): 504–14, 1992.

REVIEWS AND COMMENTARY

1. Reviewer AR11 writes:

I read with interest this manuscript regarding giant left atrium following several previous valve repairs/replacements. This manuscript illustrates very well the difficulties experienced in the management of several problems: 1) giant left atrium, 2) reoperative valvular procedures, and 3) management of end-stage cardiac dysfunction. Although the result of this report was less than optimal, I think it is not totally unexpected for a patient as sick as the one presented herein. The authors present a well thought-out approach to their problem and illustrate the procedure well. A good review of the literature is also provided for future reference.

Authors' Response by Giulio Rizzoli, MD:

Our operation was unsuccessful not because of limitations of the technique (which can optimally deal with the pathology) nor because of faulty heart function. In fact the heart function was good and the heart was optimally protected by applying the same preservation procedures as in heart transplantation. Complexity of the operation cannot be blamed as it is a very simple atrial operation. In this particular case we think the severity of adhesions, the very long "on pump" dissection and the reduced liver function from chronic relapsing CHF were the dominant incremental risk factors.

2. Reviewer JZ37 writes:

I found the case fascinating and informative. Autotransplantation in this case may have been appropriate, but I'm unsure as to the reason the thoracic surgeon found stenting the left mainstem not feasible. Whether or not a stent would have improved the patient is academic now, but could have eased the patient from the ventilator the last time and possibly brought down the P.A. pressures with improved ventilation. I am not sure that the autotransplantation was necessary as well since one might have improved the patient with a lesser plication-like operation directed at areas of concern such as the P.A., bronchus, and the posterior-inferior extension behind the L.V. thus avoiding the quite vascular "early" adhesions found due to the prior left anterior thoracotomy.

Authors' Response by Giulio Rizzoli, MD:

This was also our desire. On the contrary our consultant

felt it was too dangerous to place a stent in a completely obliterated malacic left main bronchus. He feared that, in face of a continuing external pressure from the left atrium, the stent will eventually perforate within this chamber. As we stated in the manuscript, our surgical group was split between those favoring a limited operation involving an intraatrial plication of the Kawazoe's type and those favoring autotransplantation. One of the authors strongly supported the Kawazoe plication. The other author supported an opposite opinion. Retrospectively it is obvious the choice was not correct. It is hazardous to extrapolate on a single case. Nonetheless we blame the unusual severity of the adhesions in this particular patient as the main cause of our failure and remain enthusiastic of the autotransplantation procedure. We seriously doubt an intraatrial procedure could have reduced effectively an atrium of this size with walls firmly adherent to the surrounding structures. We stress that autotransplantation of a heart with good ventricular function allows optimal heart preservation during the procedure and a superior exposure.

© 1998 Forum Multimedia Publishing