

## Mirror Image Gerbode or Partial Atrioventricular Canal Defect?

Cem Arıturk, Ass.Prof.<sup>1</sup>, Ahmet Umit Gullu, Ass.Prof.<sup>1</sup>, Sahin Senay, Ass.Prof.<sup>1</sup>, Eyup Murat Okten, M.D.<sup>2</sup>, Fevzi Toraman, Prof.<sup>3</sup>, E. Hasan Karabulut, Prof.<sup>1</sup>, Letisya Melengic, M.S.<sup>1</sup>, Cem Alhan, Prof.<sup>1</sup>

Gerbode defect, that can accurately be treated surgical repair, is defined as a true communication between left ventricle and right atrium. A 74-year-old woman with a worsening history of orthopnea and peripheral edema was hospitalised. A communication between right atrium and left ventricle was diagnosed using transeosophageal echocardiography. The defect was repaired and mitral valve was replaced with a biologic valve. It would be better to tailor surgical strategy for each case with atrioventricular canal defect after preoperative transeosophageal echocardiography and peroperative direct sight.

Key words: 1. Atrioventricular septal defect  
2. Right atrium  
3. Left ventricle  
4. Mitral valve

### CASE REPORT

The definition of a Gerbode defect, according to the Society of Thoracic Surgeons Congenital Heart Nomenclature and Database Project is true left ventricular (LV) to right atrial (RA) communication [1]. However, no sources provide a definition encompassing congenital right ventricular (RV) to left atrial (LA) communication. Zacharkiw and Stimpson [2] recently described this pathology as a 'mirror-image Gerbode defect' in a patient following atrioventricular (AV) septal defect repair. In the present study, the case of a congenital LA-RV shunt in an adult is presented and the classification of such defects is discussed.

A 74-year-old woman was referred to our center with a worsening history of orthopnea, paroxysmal nocturnal dyspnea, and peripheral edema. Jugular venous distention and pansystolic murmur were found in the physical examination.

Transthoracic echocardiography revealed LV hypertrophy, a dilated left atrium, severe mitral valve insufficiency, and pulmonary hypertension (60 mmHg). Elective surgery was planned for mitral valve intervention.

A careful review of the preoperative transeosophageal echocardiography (TEE) revealed a clear jet across a small defect between the RV and LA (Figs. 1, 2). The pericardium was entered through a midsternal incision and a patch was fixed using glutaraldehyde. Following the establishment of cardiopulmonary bypass (CPB), the heart was arrested via aortic cross-clamping and cold-blood cardioplegia. The mitral valve was exposed through left atriotomy. A vertical cleft separating the anterior leaflet into two hemileaflets was observed on the mitral valve (Fig. 3). A non-traumatic congenital anatomical defect (12 mm) was noted just superior to the anterior leaflet. Aspirator-guided inspection showed that the defect was located between the LA and the RV (Fig. 4). The seg-

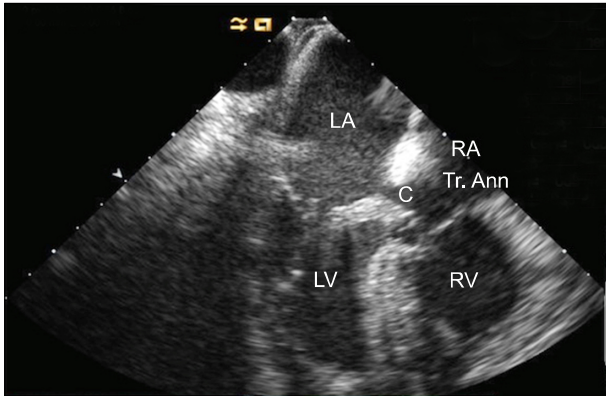
<sup>1</sup>Department of Cardiovascular Surgery, Acıbadem University, <sup>2</sup>Department of Cardiovascular Surgery, Acıbadem Kadikoy Hospital, <sup>3</sup>Department of Anesthesiology, Acıbadem University

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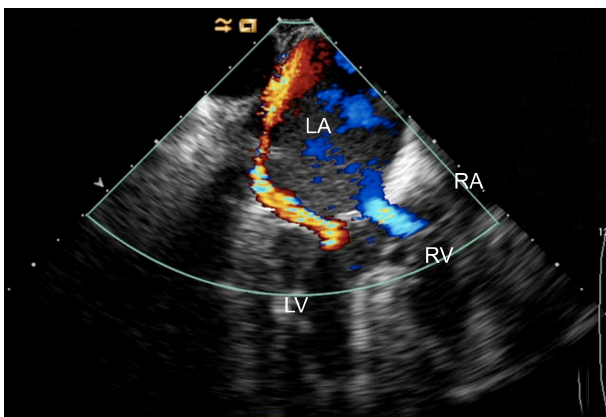
Corresponding author: Cem Arıturk, Department of Cardiovascular Surgery, Acıbadem Kadikoy Hospital, Acıbadem, Kadikoy-Istanbul 34718, Turkey (Tel) 90-505-3146824 (Fax) 90-216-5444580 (E-mail) [cemariturk.kvc@gmail.com](mailto:cemariturk.kvc@gmail.com)

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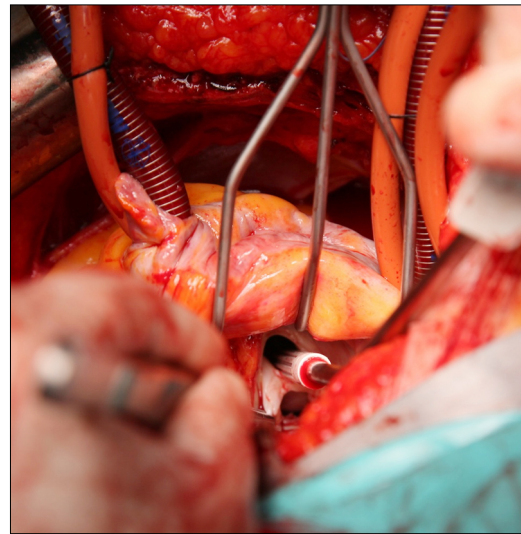


**Fig. 1.** Transesophageal echocardiographic image showing a defect between the left atrium and right ventricle. LA, Left atrium; LV, left ventricle; RV, right ventricle; RA, right atrium; C, communication; Tr. Ann, Tricuspid valve annulus.

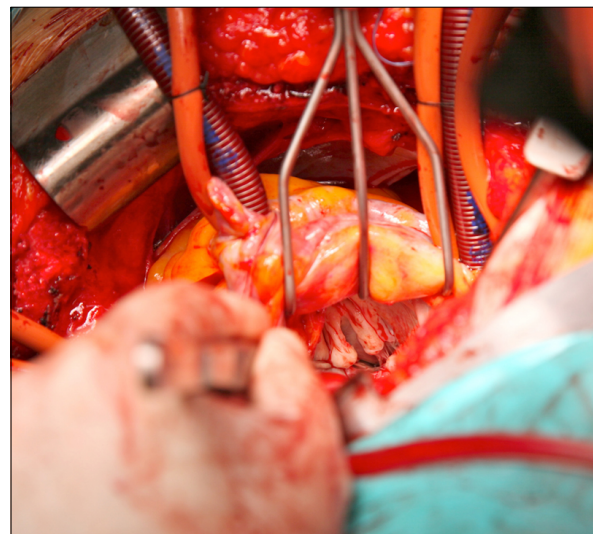


**Fig. 2.** A color Doppler image shows the jet flow. LA, left atrium; LV, left ventricular; RV, right ventricular; RA, right atrium.

mental anatomy of the posterior leaflet was normal. The saline test revealed severe mitral insufficiency at both the central and commissural locations. The cleft was closed without tension after resection of the abnormal chordae attached to its edges. The LA-RV shunt was closed using a pericardial patch. Direct suture was not technically possible due to the fibrous reaction of the edges of the cleft with an area lacking valvular tissue. Instead, the edges of the cleft were resected and the anterior leaflet of the mitral valve was reconstructed using an autologous pericardial patch. However, the saline test revealed regurgitation at both commissures. The patch was then replaced with a 27-mm porcine bioprosthetic valve (Biocor; St.



**Fig. 3.** An intraoperative view of the hole between the left atrium and right ventricle, located just above the cleft of the anterior mitral leaflet.



**Fig. 4.** Surgical exposure of the cleft separating the anterior mitral leaflet into two hemileaflets.

Jude Inc., St. Paul, MN, USA) which was implanted in a supra-annular fashion with the use of interrupted, pledgeted, 2-0 everting mattress sutures. Rewarming was initiated, the atriotomy was closed, the heart was de-aired, and the cross-clamp was removed. After rewarming, CPB was terminated without any difficulty or inotropic support. A TEE examination revealed a normally functioning prosthetic mitral valve and no shunt.

## DISCUSSION

The AV junctions are the area of the heart where the atrial myocardium is inserted into the base of the ventricular mass. Very limited contact is present between the LA myocardium and the muscular ventricular septum. Partial AV septal defects are malformations with two AV valve orifices and no interventricular communication, whereas complete AV septal defects have a common AV valve orifice and extensive interventricular communication [3].

LV to RA atrial communications, known as Gerbode defects and LV-RA shunts, are encountered from time to time and are caused by surgical mishaps, trauma, and endocarditis. However, we could find only three cases of mirror-image Gerbode defects (LA-RV shunts) that have been reported to date in the literature [2,4-6]. Of those cases, only one was congenital, while in the other cases, the defect emerged after the repair of an AV septum. The previously reported congenital case was a 39-year-old woman with a common AV junction and partially separated right and left AV orifices, and the shunt was exclusively from the LA to the RV due to overriding of the left AV valve [6]. However, in our case, each atrium was connected to its own ventricle through separate leaflets. Additionally, a cleft in the anterior mitral leaflet existed, which may have been linked developmentally, on the basis of different degrees of failure of fusion of the AV endocardial cushions.

A literature review demonstrated that it is virtually impossible to categorize the spectrum of AV septal defects into satisfactory and noncontroversial subgroups with regard to pa-

tients such as ours. We believe that diagnosis and surgical treatment strategy will become easier if case-specific morphologic and functional variables are analyzed.

## CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

## REFERENCES

1. Jacobs JP, Burke RP, Quintessenza JA, Mavroudis C. *Congenital Heart Surgery Nomenclature and Database Project: ventricular septal defect*. Ann Thorac Surg 2000;69(4 Suppl): S25-35.
2. Zacharkiw LA, Stimpson HL. *Mirror-image Gerbode defect?* Eur J Echocardiogr 2011;12:627.
3. Kirklin JW, Barratt-Boyes BG. *Cardiac surgery: morphology, diagnostic criteria, natural history, techniques, results, and indications*. 2nd ed. New York (NY): Churchill Livingstone; 1993.
4. Chirillo F, Totis O, Cuzzato V. *Left-atrium-to-right-ventricle shunting after surgery for atrioventricular septal defect: an unusual sequel detected by transesophageal echocardiography*. Int J Cardiol 1992;37:253-6.
5. Mohapatra S, Minhas HS, Virmani S, Mishra BB, Mukherjee K, Banerjee A. *Acquired left atrial-to-right ventricular shunt with mitral valve incompetence: a rare sequela after repair of atrioventricular septal defect*. Tex Heart Inst J 2009;36: 69-71.
6. Delgado C, Bonnin O, Barturen F. *An unusual tethering of the bridging leaflets in atrioventricular septal defect producing a communication from left atrium to right ventricle*. Cardiol Young 1999;9:84-7.