

Gerbode Defect, Importance of Left Ventricle-Right Atrial Shunt

Gerbode Defekt, Sol Ventrikül-Sağ Atrium Şantı Önemi

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ABSTRACT Gerbode defect is an abnormal shunt between the left ventricle and right atrium (LV-RA). The shunt can be either congenital or acquired. The pathophysiology is the injury of the atrioventricular septum, thus it leads abnormal shunting of the blood. Although it is a rare anomaly, the development of technology in the pediatric cardiology and cardiovascular surgery era or the increased number of adult congenital heart disease patients, when suspected one should consider the left ventricle to right atrial shunt.

Keywords: Heart septal defects, ventricular; echocardiography

ÖZET Gerbode defekt sol ventrikül sağ atrium (LV-RA) arasında oluşan şanttır. Doğumsal veya edinsel olabilir. Patofizyolojiden sorumlu tutulan atrioventriküler septum hasarı sonucu kanın anormal olarak sol ventrikül sağ atrium arasında şantı gerçekleşir. Nadir bir anomali olmasına rağmen, çocuk kardiyojide görüntüleme teknolojisindeki ve kardiyovasküler cerrahideki ilerlemeler, artmış erişkin dönem konjenital kalp hastalığı olan vakalar nedeniyle, LV-RA şant düşünülmelidir.

Anahtar Kelimeler: Kalp septum kusurları, ventriküler; ekokardiyografi

Left ventricle (LV) right atrium (RA) connection is a rare connection anomaly. Congenital LV-RA shunt was first declared in an autopsy report.¹ In 1949 Perry reviewed five cases from the literature, by adding another patient to the literature, he also described the anatomical variation.² In 1957, Kirby successfully closed LV-RA shunt during ventricular septal defect (VSD) operation.³ Gerbode successfully closed this defect in five patients, and so named it as “Gerbode defect” (1958).⁴ It accounts for <1% of all congenital heart defects.⁵ The etiology of LV-RA shunt is primarily congenital, but can be acquired in nature as well. In some studies, the incidence of LV-RA shunt is 8-15% in VSD patients. It

is clear that some defects develop from membranous VSDs as a result of structural changes that form an aneurysmal pouch with tricuspid valve tissue and occur during spontaneous closure.⁶ Wu et al. reported 8.1% of left ventricular-to-right atrial shunt, while Ramaciotti et al. reported 15%.^{7,8} In recent years the authors also mentioned acquired LV-RA shunt.⁹ With the advances of noninvasive diagnostic techniques including echocardiography, computed tomography, and magnetic resonance imaging, the acquired shunts as a result of infective endocarditis (IE), trauma, valve replacement, or myocardial infarction have been increasingly discovered. Nevertheless, the accurate prevalence remains unknown.⁶

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This article provides a review of LV-RA shunt by means of anatomy, physiology, symptoms, diagnosis and treatment modalities.

ANATOMY

The anatomical relation of Gerbode defect was classified by many authors. Riemenschneider has classified the defect into two: direct and indirect types.¹⁰ On echocardiographic examination of direct (true) defects, the blood in the left ventricle goes through the membranous septum, which is above the tricuspid valve. In patients with perimembranous ventricular septal defect, the shunt is from left to right ventricle passes through the tricuspid valve into the right atrium. This occurs below the tricuspid valve, which is called as indirect LV-RA shunt (Figure 1).¹¹

Another classification is that supravalyvular and infravalvular defects. One-third of the defects are supravalyvular defects, and the remaining two-thirds are infravalvular defects.¹¹ In 1962, Sakakibara and Konno described another type of defect known as intermediate type.¹² Taskesen et al. described the defects as supravalyvular (type 1), and infravalvular (type 2 and 3) (Figure 1) Type 2 and 3 defects have anomalies of tricuspid valve, such as cleft.¹³ Supravalyvular defects are located in the atrioventricular membranous septum. Because of its location, it causes a direct shunt between RA and LV. Infravalvular defects are located below the septal leaflet. Some infravalvular defects are proposed to develop from aneurysmal pouches that result from perimembranous VSDs spontaneous closure. Not only formation of tricuspid aneurysmal pouch, but also tricuspid valve perforation, fenestration, comis-

sural insufficiency and also tricuspid valve cleft can cause LV-RA shunt.⁷

Wu et al. found that integration of anterior leaflet tissue was strongly associated with LV-RA shunting. Less frequently, the leaflet can be malformed, perforated or in case of cleft, also LV-RA shunt can be observed.⁷

PATHOPHYSIOLOGY

In Gerbode defect, shunt occurs from LV to RA. Due to shunt, the blood passes through right ventricle, and as a result, the right heart chambers, first right atrium, and then right ventricle dilates, which is a decisive finding of LV-RA shunt.¹¹ Because the right atrial pressure increases, the physiology interferes with pulmonary arterial hypertension. This can be distinguished from echocardiographic and catheter angiographic studies. On the other hand, if the shunt is large, due to overflow, the left heart becomes enlarged.¹⁴

Acquired defects are subcategorized as acquired iatrogenic Gerbode defects (AIGD) and acquired noniatrogenic Gerbode defects (ANIGD). The most common reason of AIGD is the LV-RA shunt after cardiac surgery.

A recent study has shown that acquired LV-RA shunts are common in males (68%) and the average age is 49 years (2 to 86 years).¹⁵ A previous cardiac operation such as mitral, aortic valve replacement surgery or VSD operations are the most common causes of acquired LV-RA shunt.¹⁶⁻¹⁸ After valve surgery, one should consider LV-RA shunt in case of acute decompensation concomitant with arterial desaturation. After primum ASD and mitral cleft repair, LV-RA shunt can be seen as a complication.¹⁹ Some

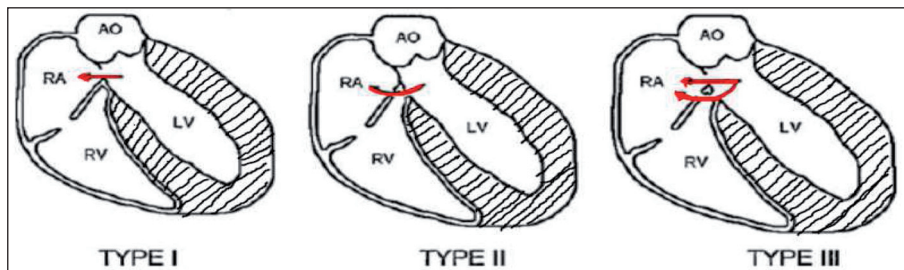


FIGURE 1: The classification of LV-RA shunt. The most common accompanying lesion of congenital LV-RA shunt is ASD in 1/3 patients.⁶

surgical techniques have been suggested to prevent this complication.²⁰ In patients having acquired LV-RA shunt, reoperation is also an important risk factor because of the probability of hemodynamic instability and/or complete AV block.²¹

The second most important cause of acquired LV-RA shunt is infective endocarditis.²¹ Endocarditis has been shown to cause LV-RA shunt by reopening a congenital defect, widening a small, insignificant shunt or by destructive perforation of the septum. It has been reported that LV-RA shunts in association with VSDs increases the risk of endocarditis (58 per 10,000 patient-years) in comparison to typical VSDs or mitral regurgitation

The defect caused by infective endocarditis can be type 1 (65%) and type 2 (26%); type 3 defect is rare. Staphylococcus (40%) and Streptococcus (40%) species are the usual cause.²¹

In the elderly, myocardial infarction (MI), especially inferior MI is the other cause of acquired LV-RA shunt. Its mortality rate is 80%. Infarctions with VSD have a worse outcome when the infarction is located inferiorly.²²

The other reason for acquired LV-RA shunt is trauma, such as blunt car accident, or penetrating injury. Occasionally, other accompanying injuries occur with LV-RA shunt. Pure LV-RA shunts are less seen due to trauma.²³ There is a case of acquired LV-RA shunt, whose initial finding was complete heart block after a motorcycle accident and operated without complication.²⁴

Iatrogenic defects are another important cause of acquired LV-RA shunts. There are cases having been reported during catheterization, operation or electrophysiologic ablation procedure.²⁵⁻²⁷

SYMPTOMS

In LV-RA shunt, the symptoms vary from asymptomatic to severe. It depends on the volume and duration of the shunt. Dyspnea occurs due to pulmonary over circulation. Right heart failure symptoms such as weakness, fatigue, shortness of breath, and lower extremity edema are also the other symptoms.¹⁴ These symptoms are also seen in the clinical condition of pulmonary arterial hypertension (PAH). One

should differentiate PAH vs LV-RA shunt by using transthoracic echocardiography (TTE), and transesophageal echocardiography (TEE).

PHYSICAL EXAMINATION

The murmur is similar to that of VSDs, pansystolic, grade 3-4. It can be difficult to distinguish from VSDs murmur. Vogelpoel reported that VSDs murmur has a higher frequency and becomes softer during inspiration.²⁸

Right heart failure findings such as peripheral edema and elevated jugular venous pressure are also common.

If the shunt is large and develops acutely, basal crepitant rales can be auscultated due to pulmonary over circulation.

DIAGNOSIS

TRANSTHORACIC ECHOCARDIOGRAPHY

The basic transducer positions are parasternal short-axis, apical short axis, and subcostal views. Color flow Doppler is valuable for revealing high-velocity systolic flow (>4 m/s) between LV and RA. The characteristic stream must be distinguished from the other conditions such as ruptured sinus of Valsalva aneurysms, endocardial cushion defects, VSD, and Tricuspid Regurgitation. There are some echocardiographic clues to suggest Gerbode defect as follows (Figure 2);¹¹

1) atypical jet direction; 2) lack of ventricular septal flattening; 3) persistent low velocity shunt flow into systole; 4) no right ventricular hypertrophy; 5) normal diastolic pulmonary artery pressure.

Differences in the timing of the shunt help to distinguish Gerbode defect from sinus valsalva rupture. Gerbode defect produces left to right shunt during systole, while ruptured of Valsalva produce diastolic shunt (Figure 3).¹¹

Another important diagnosis to distinguish is pulmonary arterial hypertension (PAH). If the systolic flow misinterpreted as tricuspid regurgitation, severe PAH will be diagnosed incorrectly.¹¹ So one should do the differential diagnosis of pulmonary arterial hypertension by means of tricuspid regurgitation.

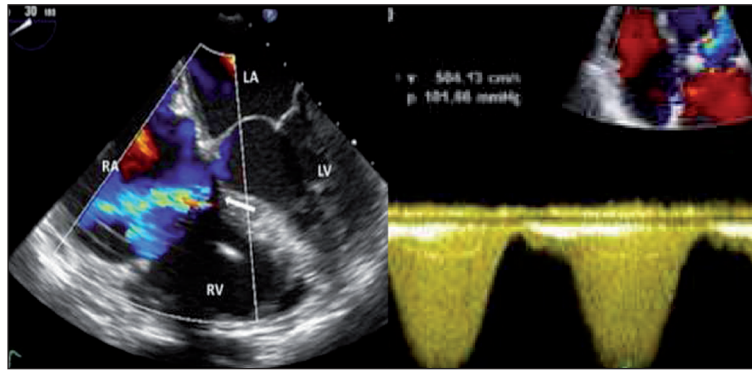


FIGURE 2: LV-RA color flow and CW Doppler of the shunt.

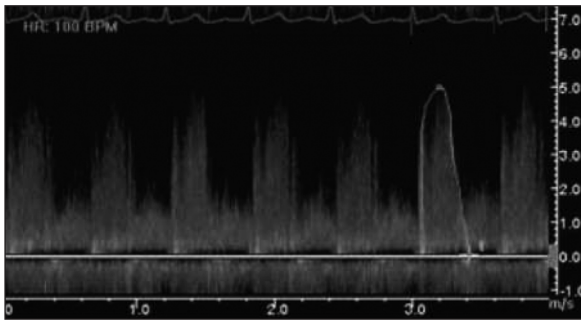


FIGURE 3: Diastolic flow of ruptured sinus valve.

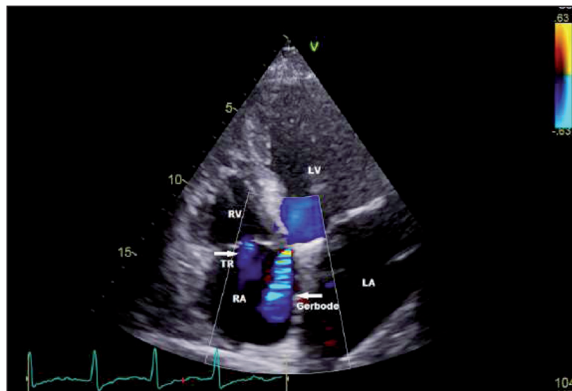


FIGURE 4: LV-RA shunt and tricuspid regurgitation.

To distinguish PAH from Gerbode defect jet flow, the normal diastolic pulmonary arterial pressure in Gerbode defect can be used (Figure 4).

TRANSESOPHAGEAL ECHOCARDIOGRAPHY

Two-dimensional TEE has limitations, because of its visualization of the anatomic location of the defect and

its relationship with the other structures. Nevertheless, real-time three-dimensional (RT 3D) echocardiography is more appropriate to detect the structural abnormalities. The RT 3D TEE is used not only in diagnosis but also during percutaneous treatment (Figure 5).¹¹

MAGNETIC RESONANCE IMAGING

Cardiac magnetic resonance imaging (MRI) also gives detailed anatomical and physiological information about the defect, such as anatomy, heart volumes, quantification of shunt flow. Although it has advantages, high cost and limited availability are the disadvantages.¹³

CARDIAC CATHETERIZATION

Cardiac catheterization is the gold standard to assess the hemodynamic situation. It is the best method to distinguish Gerbode defect from PAH. The LV-RA shunt can be confirmed by left ventriculography.

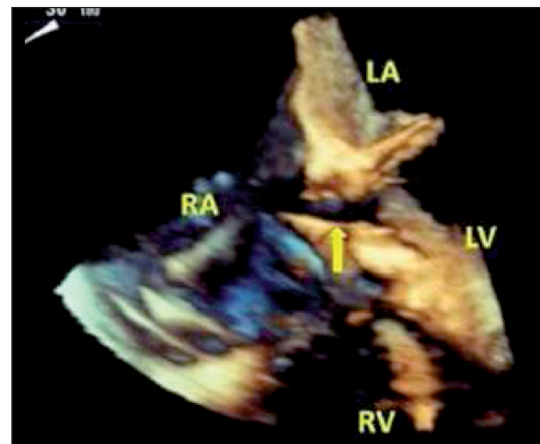


FIGURE 5: Real 3D TEE of LV-RA shunt.

Opacification of right atrium prior to the right ventricle is the key. There is an 20% oxygen step-up in RA, compared to the SVC.¹⁴

TREATMENT

Treatment depends on the symptoms, other structural heart defects, and comorbidities. Small or asymptomatic defects can be managed conservatively.²⁸ Some authors suggest that asymptomatic patients with insignificant shunt should be under close follow-up rather than surgery.²⁹ Nevertheless, the others suggest that all LV-RA defects should be repaired immediately when the diagnosis is correct, that's because of increased infective endocarditis risk.³⁰

During surgery, a patch repair is often performed on the right atrial side. There are some other surgical techniques, such as patch repair with sutures from the ventricular side of the tricuspid valve through the leaflets or Dacron patch closure with septal leaflet reimplantation onto the patch, and so on.^{20,31,32}

The other closure technique is catheter based treatment. Previous devices have been used for this reason. There are case reports in the literature. Amplatzer occluder device, PDA duct occluder, VSD coil are the devices having been used for closure of Gerbode defect.^{10,28} Among these, the Amplatzer occluder device is a mainstay in treatment, because of its less radial force (Figure 6).¹⁰

Acquired LV-RA shunts may be associated with infective endocarditis, congestive heart failure, valvular leaflet perforation, complete atrioventricular block. That's because the surgical operation should be preferred despite percutaneous devices.²²

CONCLUSION

The left ventricular-right atrial shunt is a very rare anomaly. It is generally associated with the other congenital heart diseases, or in acquired forms with other comorbidities, such as infective endocarditis. A Gerbode defect should be suspected when an unusually

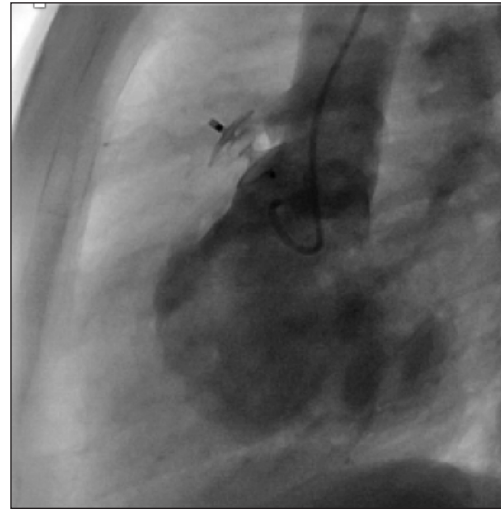


FIGURE 6: Percutaneous closure of LV-RA shunt with Amplatzer occluder device.

dilated right atrium or high Doppler gradient between the left ventricle and the right atrium is seen on echocardiography. Spontaneous closure of these defects is very rare. Surgical closure is safe, but in recent years interventional therapy has become the alternative way of treatment. After treatment, excellent results can be seen.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

This study is entirely author's own work and no other author contribution.

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