Communication anomaly among left ventricle and right atrium; Gerbode defect.

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Abstract

Gerbode defect is a rare cardiovascular defect occurring as a septal abnormality among the left ventricle and right atrium. Acquired Gerbode defect had surpassed the genetic defect rate with increasing prevalence in old individuals. It can also occur as a rare complication or secondary condition due to repeated replacement of heart valves. The overwhelming majority of cases are thought to be caused by iatrogenic injury, endocarditis, myocardial infarction in the right coronary artery distribution, or blunt cardiac trauma. Dyspnea, congestive heart failure, orthopnea, and pedal edema are prevalent symptoms experienced after the mitral or aortic valve replacement procedure. Transesophageal Echocardiography (TEE), Transoesophageal Echocardiogram (TOE), and Echocardiography (ECG) are used along with Doppler for the diagnosis. Elevated atrial volumes, irregular heartbeats, and atrial fibrillation are the identifying factors for the septal defect. It is a high-risk condition that requires immediate care, such as surgical management. The pericardial patch is opted prevalently during the surgical treatment of defects.

Keywords: Gerbode defect, Left Ventricle Right Atrium communication, Mitral Valve Replacement, Aortic Valve Replacement, Mitral Stenosis.

Introduction

Rare ventricular septal abnormalities known as the Gerbode defect include Left Ventricular (LV) to Right Atrial (RA) connections [1]. Thurman J. originally wrote about this defect in 1838. However, when Professor Frank Gerbode documented a pioneering case series that included five successful operative repairs, it was given his name [2]. Abnormal communication between the left and right atrium occurs, which can be due to several causative factors such as cardiovascular diseases like endocarditis, myocardial infarction, and stroke or as a result of genetic abnormalities [3]. According to Gerbode et al., the

flaws in the communication of left and right ventricles are typically congenital but can be acquired as well in rare cases.

There are two types: either an absence of the atrioventricular membranous septum or shunting that first occurs due to a ventricular septal defect, with atrial shunting resulting from an error that affects the tricuspid valve's septal leaflet [4].

Cause

Congenital defects are relatively rare (0.08%) and can be caused by a membranous Ventricular Septal Defect (VSD) and an anomaly in the leaflet of the tricuspid valve, or they

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might develop indirectly (1). Gerbode abnormalities are uncommon, and while some instances' pathophysiology has been linked to sequence variants in the NK2 Homeobox 2 (NKX2-5), GATA4, and T-box transcription factor 5 (TBX5) genes, the bulk of cases' causes are still unknown [5].

The condition was initially thought to be a rare genetic defect, but recent research indicates that acquired defects are now more common than congenital ones. The overwhelming majority of cases are thought to be caused by iatrogenic injury, endocarditis, myocardial infarction in the right coronary artery distribution, or blunt cardiac trauma [6].

Adults may infrequently experience acquired LV-RA shunts due to a defect in the ventricular septum's upper section [7]. It was also presented as a rare complication or secondary condition due to repeated replacement of heart valves [8]. There have been reported presentations of communication errors between valves due to the injury that occurred during surgery, which caused a shortcoming in the membrane of the atrioventricular septum [9]. A defect in the membrane interventricular septum, which is located below the crista supraventricularis, is the cause of both types of LV-RA connections. Earlier cardiac surgical procedures (mitral valve replacement, aortic valve replacement, VSD-ASD (atrial septal defect) repair, and percutaneous cardiac interventions (AV node ablation, endomyocardial biopsy, and tricuspid annuloplasty), which account for the majority of the rise in over the past 20 years, are the two primary contributors of acquired iatrogenic Gerbode defects.

Anatomy

The septal leaflet of the TV typically lies 5 to 10 mm higher towards the apex than the anterior leaflet of the mitral valve, considering the annulus of the tricuspid valve occupies a position more apically than the mitral annulus. Due to this structural configuration, the supravalvular (also known as the atrioventricular) and intra-valvular (also known as the interventricular) sections of the membrane ventricular septum are separated by the septal leaflet. A direct LV-RA connection is the outcome of a supravalvular section defect. Connectivity between the right ventricle and the left ventricle would typically emerge from a peri-membranous ventricular septal defect in the intra-valvular region [10].

Symptoms

When there are Gerbode deficiencies, the RA experiences pressure as well as volume overexertion, which is accompanied by symptoms such as dyspnea and heart failure. Physical examination reveals a noticeable pan systolic murmuring at the left sternal border [11]. An LV-RA shunt can be identified physically by a loud, harsh, pan-systolic murmur that is frequently accompanied by a thrill across the left side sternal border. However, a diagnosis cannot typically be made only on the grounds of the results of a physical examination.

Edema and shortness of breath or dyspnea are the prevalent physical symptoms observed among patients with communication defects in the chambers of the heart, mainly the left ventricle and right atrium. Heart murmur is also identified as a symptom after the mitral or aortic valve replacement surgery, such as pansystolic murmur as well as holosystolic was observed. Edema in lower limbs, pedal edema, asities, and congestive heart failure or endocarditis are also listed as symptoms [12].

Diagnosis

The preferred diagnostic method is TEE. The flow of color Doppler will show the presence of, while attenuated throughout diastole, a high-velocity systolic jet with flow directed toward the Right Atrium (RA). The advancement of innovative methods for imaging has increased the reliability in the identification of acquired Gerbode defects, especially those that arise from adverse effects of cardiac procedures and percutaneous coronary interventions involving the interventricular septum or adjacent structures, such as cardiac magnetic resonance and multidimensional Transesophageal Cardiac Echocardiography (TOE).

It's crucial to differentiate between tricuspid regurgitation and LV-RA communication. Unusual jet direction, continuous shunt flow into diastole, insufficient ventricular septal flattening, lack in right hypertrophy of the ventricular walls, and normal diastolic pulmonary artery pressure as determined by the pulmonic regurgitant velocity are some of the most significant echocardiographic clues that point to the presence of the Gerbode defect [13]. Elevated atrial volumes can cause structural alterations that lead to irregular heartbeats, including atrial fibrillation [14].

Treatment plan

The reduction of vascular resistance in the system, an increase in the output of the heart, and a decrease in lung congestion constitute the hemodynamic objectives for the defect [15].

Physiology

As a consequence of the substantial blood pressure gradient that prevails among these heart chambers, switching from LV to RA happens physiologically. The passage of blood into the Right Atrium (RA) is enhanced as a result of tunneling, which enlarges the right side of the heart compartment.

Management of disease

In patients with these abnormalities, maintaining native or temporarily assisting atrioventricular pumping is crucial to promoting appropriate LV filling, volume of stroke, and outward stream in the presence of significant regurgitant circulation and heart failure-related symptoms.

An Amplatzer occluder apparatus can be employed in the cardiovascular interventional unit to provide definitive treatment [16]. Alternately, an extremely risky invasive surgical alternative known as the commando method, which combines mitral and aortic valve replacement alongside intervalvular fibrous repairs to the body, could be performed [17].

Mitral valve replacement

We conducted a literature-based analysis of case reports research to identify the characteristics, clinical impact,

symptoms, diagnostic procedure, management, intervention for the defect. The majority of patients diagnosed with Gerbode defect are older adults except for two individuals aged 23 and 25. A twenty-five-year-old male patient with arterial septal defect and cleft mitral valve had undergone a surgical procedure for the treatment of the condition at 16 years. CV-LA communication deficit was reported after nine years of surgery and diagnosed with the help of ECG and color flow. The second patient was 23 years old with a history of mitral and tricuspid valve replacement due to iatrogenic factors. Hematuria, hemolysis, and pan systolic murmur were reported after three months of surgery. Surgical management of defects was employed for optimal outcomes [18]. Dyspnea

was one of the prevalent symptoms among all the patients, along with murmur, chest pain, and edema. Congestive heart failure and orthopnea were typically present in 4 of 16 patients reported in this study Figure 1-4.

Age

Occurrence of Gerbode defect was more prevalent in the population above 40 years, with almost 50% of patients in the age bracket of 51-60 years and above 70 years. The maximum age of patients suffering from LV-RA defect among communication was 81 years male, as reported by Sunderlandet al. After five years of surgery for aortic valve replacement. 2 patients, making up 11% of research patients reported in this

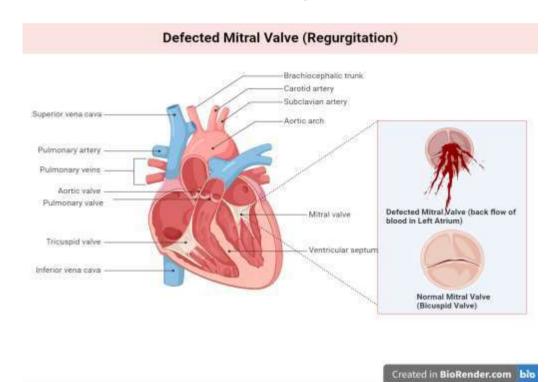


Figure 1. Mitral Valve Regurgitation.

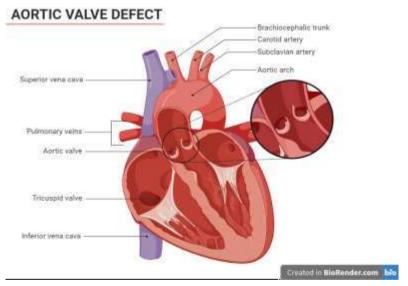


Figure 2. Aortic Valve Defect (AVD).

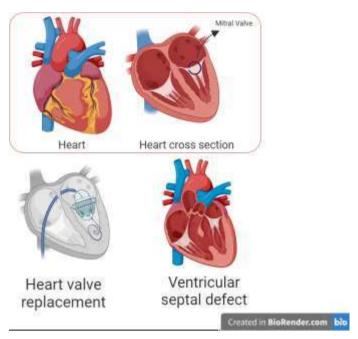


Figure 3. Ventricular Septal Defect (VSD), Heart Valve Replacement.

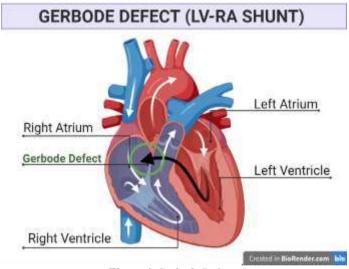


Figure 4. Gerbode Defect.

study, are in their twenties, indicating infrequent occurrence of acquired Gerbode defect in young individuals. A study reported a case of a 30-year-old female patient with mitral valve replacement in the past with infected endocarditis diagnosed by utilizing TOE and ECG. The Gerbode defect occurring secondary to any disease is an acquired defect in cardiovascular chambers.

Diagnosis

Myocardial stenosis was the prevalent causative agent among patients with Gerbode defect. There was also an incidence of myocardial stenosis along with tricuspid regurgitation. Aortic stenosis occurred in two patients, as reported by the research authors. There was also a diagnosis of endocarditis in 3 patients. An incident of iatrogenic occurrence of Gerbode defect was also presented by the researcher [19-22].

Diagnostic method

ECG was utilized in almost all the cases of Gerbode defect diagnosis. Many studies used TOE, which stands for transoesophageal echocardiogram, for the purpose of identification of the LV-RA communication impediment in the heart of patients. TOE is conducted via the probe. The probe is inserted via the esophageal route of the patients to find a clear and precise image of the heart for a better understanding of the disease. Left ventriculography was used by Aoyagi et al. only as the patient was suffering from tricuspid regurgitation (TR). 2 case reports included the Doppler color method as a diagnostic tool for obtaining a colorful and reliable image of the thoracic cavity [22-27] (Table 1).

Causative agent

Myocardial valve replacement surgery was identified as one of the potential causative agents for communication defects

acquired in the heart after surgery between two chambers, the left ventricle and right atrium. Myocardial replacement surgery was also conducted in conjunction with tricuspid annuloplasty, which was also reported in the two patients of the research case report. Aortic Valve Replacement was reported by Fanari et al. in one patient and Sunderland et al. in two patients, whereas Dzwonczyk & Davidson reported Replacement of both aortic and mitral valve in a patient.

Symptoms

Edema and shortness of breath or dyspnea were the most frequently reported symptoms by the patients. Heart murmur of pansystolic as well as holosystolic was also observed. Edema was observed mainly in the lower extremities of the body, mainly pedal edema among three patients suffering from Gerbode defect or communication barrier between the chamber of heart LV and RA. Congestive heart failure with prominently acute presentation was also documented in the studies [28].

Type of Gerbode Defect

All the case studies reported Gerbode defect as an acquired deficit in the ventriculoarterial chamber communication.

Time duration

Patients within higher age brackets demonstrated reported heart defects within a short period of time, mainly months, such as a 77-year female presentation of the defect within two months, and the same occurrence of the brief time period was observed with the increasing age except for Sunderland et al. that reported time period of 5 years after the aortic valve surgery and history of Coronary Artery Bypass Grafting (CABG) at the age of 81 in a male patient.

Comorbidities

Pulmonary hypertension was significantly reported to be greater than other comorbidities among the patients.

Congestive hepatomegaly was present in a 51-year-old female patient, reported by Moaref et al., and Hodgkin lymphoma was present in a 54-year-old male patient suffering from acquired Gerbode defect and acute heart failure along with hemolytic anemia, acute kidney injury, and aortic stenosis. Non-metastatic cancer of the large intestine, particularly the colon (Duke's carcinoma), was presented by Collis et al. with a history of right hemicolectomy in 73-year-old male patients suffering from mitral valve regurgitation.

Management of disease

Surgical management of disease was opted by the majority of studies with methods such as pericardial patch closure, with two studies opting for interventional procedures like catheterization as a less invasive approach owing to the condition of their patient as surgery is a high-risk procedure for this disease.

Limitation and recommendation

We gathered the case reports for the rare condition of Gerbode defect characterized by LV-RA communication defect. Our main focus was the acquired Gerbode defect, as it had surpassed the genetic occurrence of the condition. It would have been better to gather and compare the genetic and acquired defect characteristics and clinical implications along with the difference in their management and intervention for treatment. Such a study would be beneficial for future perspectives, acting as contributing documentation for researchers and healthcare professionals to determine and recommend an optimal treatment plan for better outcomes. Interventional strategies utilized by researchers for the treatment of this condition, along with an assessment of overall survival rates, should also be added in future studies. A study assessing the quality of life among patients of Gerbode defect, as well as a study to determine the dietary causative agents, is advised to explore in future Table 2.

Table 1	. Age Ro	inge of	Participants.
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Age Range	No of patients n (%)	Reference			
>40	3 (18.76%)	[10],[19],[25]			
41-50	3 (18.76%)	[12], [16]			
51-60	3 (18.76%)	[1], [22], [24]			
61-70	2 (12.16%)	[26], [28]			
71<	5 (31.2%)	[10], [20], [23], [27], [28]			

Table 2. Characteristics of studies.

Study	Year	Age	Sex	Diagnosis	Diagnostic Method	Causative Agent (Surgery, disease)	Symptoms	Type of Gerbode Defect	History of surgeries	Time	Comorbidities	Management
Uwabe et al.	2022	77	F	MS, TR	ECG	MVR, TAP	Pan systolic murmur, edema	Α		2 months	Pulmonary Hypertension	Surgical pericardial patch
Rebecca et al.	2021	54	М	MS, TR9	TOE	MVR & TVR	Acute Heart Failure, weight gain, SOB, orthopnea	А	Pacemaker surgery, Surgery for MS, and tricuspid regurgitation	One month	Hodgkin lymphoma, Pulmonary Hypertension, Arterial septal defect, hemolytic anemia, Acute Kidney Injury,	pursue transcatheter device closure
Moaref et al.	2008	51	F	MS	TOE	MVR	Edema, pain in lower limbs, ascites	А	Moderate tricuspid regurgitation	1 week	Congestive hepatomegaly,	Surgical operation

Aoyagi et al.	2008	71	F	TR	ECG, left ventriculography	MVR, TAP	Heart murmur, anemia	А		8 months		Surgical operation by mattress suture & bicuspid annuloplasty
Fanari et al.	2015	50	F	AS	TOE, CT	AVR	Congestive heart failure	А		12 years		Pursue Amplatzer muscular ventricular septal defect occlude.
Amirghofran & Emaminia	2009	51	F	MS	TOE	MVR	Dyspnia, lower limb edema, ascities	A		Few weeks		Surgical management
Peñalver et al.	2017	30	F	Infective endocarditis	TOE, ECG	MVR		А				Surgical management
Paolini et al.	1993	69	F	Infective endocarditis	TOE, ECG, Doppler color	MVR	Holysytolic murmur, congestive heart failure, SOB, orthopenia	А		6 months	Pulmonary HTN	Surgical management
Miller et al.	1993	41	F	MS		MVR	congestive heart failure	A	Closed mitral commissurotomy, mitral regurgitation, tricuspid insufficiency	8 years	Pulmonary HTN	Surgical management
Usle et al.	2007	54	М	MS	Doppler color	MVR	Respiratory distress, aortic and tricuspid regurgitation	Α		2 months		Surgical management
Collis et al.	2017	73	М	MR	TTE	MVR	Dyspnnea. chest pain	Α	Right hemicolectomy,		Non-metastatic Duke's (cecal carcinoma), stroke	Surgical patch closure
Abdi et al.	2015	23	М	iatrogenic	ECG, TEE	MVR& TVR	Hematuria, hemolysis, pan systolic murmur	A		3 months	anemia	Surgical management
Dzwonczyk & davidson	1995	74	F		ECG	MVR	Holosystolic murmur, pedal edema	А			Chronic atrial fibrillation, HTN, acute MI	Surgical management
Dzwonczyk & davidson	1995	25	М	Arterial septal defect, cleft mitral valve	ECG, color flow	AVR, MVR	Holosystolic Murmur	A		9 years		Surgical management
Sunderland et al.	2020	64	М	AS, CAD, infective endocarditis	ECG	AVR	SOB, pansystolic and diastolic murmur		CABG		HTN, DM-II, Hypercholesterolemia, nephrotic syndrome, pulmonary embolism	Surgical management
Sunderland et al.	2020	81	М	AS, CAD	ECG,	AVR	Delirious, SOB, pyrexial		CABG	5 years		Surgical management

Conclusions

Mitral valve replacement procedures are conducted in cardiovascular events according to the requirement of disease treatment criteria. It has been reported that there is a greater incidence of Gerbode defect, which can be characterized as a communication barrier or hindrance among the chambers of the heart, which are the right atrium and left ventricle, due to various conditions and surgical progression. Moreover, Gerbode defect is a rare event reported scarcely in the documentation of cases. Acquired Gerbode rate occurrence had surpassed genetic Gerbode defects required surgical intervention, but it is a high-risk procedure. The pericardial patch opted for the surgical plan to fill the communication between heart chambers.

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