



Case Report

A Rare Case of Gerbode Defect and Review of Literature

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Abstract: A Gerbode defect is a very rare type of congenital anomaly with a direct communication between left ventricle and right atrium leading to Left to Right shunt. This defect was first reported by Meyer in 1857. First successful closure was done by Kerby et al using hypothermia and inflow occlusion technique. It may present as a direct or indirect type. With improvements in nomenclature and taxonomy that expanded the classification until the current modifications were in place that accounted for defect type and position with respect to the Tricuspid Valve. Now, it is broadly described as Supravalvular type and infravalvular type. The etiology is typically congenital with irregularities emerging by perforation of anterior intraventricular septum, malformation of leaflets, or widening of the commissural space. These embryological deviations subsequently permit an abnormal communication that begins the physiological processes leading to the pathology. Shunt depends on the size of the defect and PVR. Shunt flow begins in utero because of the obligatory difference in the systolic pressure between LV and RA. RA accepts shunt with less or no elevation of pressures due to distensibility. We report a 8 year old female child who presented with dyspnea on exertion and recurrent respiratory infections whose echocardiography confirmed a direct type (supravalvular) Gerbode defect. This was confirmed by cath study and during surgery. She underwent successful suture closure of the shunt using a Goretex patch. This is a case of direct type Gerbode defect doing well after surgery.

Keywords: Gerbode Defect, Congenital Anomaly, Left to Right Shunt, Direct, Indirect, Ventricular Septal Defect, Tricuspid Valve

1. Introduction

A Gerbode defect is a type of acyanotic congenital heart disease with shunt between left ventricle and right atrium. This defect was first reported by Meyer in 1857 [1]. First successful closure was done by Kerby et al using hypothermia and inflow occlusion technique [2]. Dr. Gerbode described the first successful series of patients operated on with a Left Ventricle-Right Atrium shunt in 1958 [3]. Reimenschneider and Moss have described two types of the communication based on the insertion of septal leaflet of Tricuspid valve, which divides the membranous septum into interventricular and atrioventricular parts [4]. According to STS congenital

heart nomenclature [5] and database project the definition of Gerbode defect is a true Left Ventricle-Right Atrium communication.

There are two types known, a direct and an indirect type as reported by Gerbode et al. The direct type of Gerbode defect is rare with a direct communication between Left Ventricle to Right Atrium [4]. It is sometimes referred to as supravalvular defect [Figure 4]. In Indirect type, there is a perimembranous VSD and a defect in Tricuspid valve, the shunt is from left ventricle to right ventricle and then through the Tricuspid valve into Right atrium [4]. This communication occurs below

the Tricuspid valve [Figure 4]. In both defects, left ventricular outflow tract to right atrium communication allows for shunting of blood to the right atrium during systole. If this communication is large, it can lead to volume overload and chamber enlargement [6].

2. Case Report

A 8 year old child was referred to the cardiologist by a local physician due to increased symptoms of dyspnea and respiratory tract infections. She was born as a second child to a couple of non consanguineous marriage with no delay in attaining milestones. Other sibling was normal.

On examination, she had left precordial bulge. Apex beat was heard in left 4th intercostal space, mid clavicular line. There is a Grade 4/6 pan systolic murmur heard over the left sternal border with no signs of pulmonary hypertension or heart failure. An ECG revealed sinus rhythm with RA enlargement. X-ray showed Right Atrial, Right Ventricular enlargement [Figure 3]. Trans thoracic echocardiogram revealed a non dilated Left Ventricle, but dilated Right Atrium, Right Ventricle and a 0.8 cms perimembranous Ventricular septal defect with left to right shunt directing the flow from Left Ventricle into Right Atrium [Figure 1]. A LV angiogram was done which showed Gerbode defect with VSD aneurysm. Opacification of Right Atrium in one cardiac cycle followed by RV opacification, suggestive of shunt from LV to RA. She was taken up for shunt closure. Under cardiopulmonary bypass, midian sternotomy and Right atriotomy, she underwent successful suture closure of the shunt using a Goretex patch. Associated PDA was found which was ligated. The echocardiographic findings were confirmed intra operatively [Figure 2]. No residual shunt seen. The patient had complete heart block for which PPI was placed. At 3 years follow up, patient is doing well.

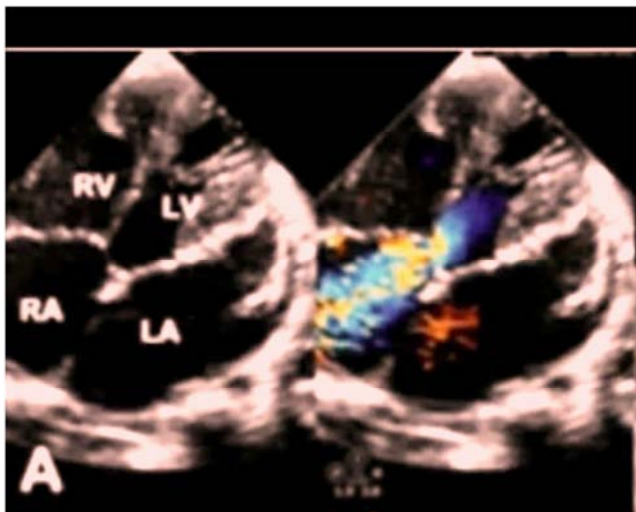


Figure 1. Echocardiogram showing the shunt from Left Ventricle (LV) to Right Atrium (RA).



Figure 2. Intraoperative picture showing the defect and its surrounding structures.

3. Result

Patient was extubated in 5 hours after surgery and shifted out of ICU on post operative day 2. She was discharged in 4 days. The patient had an uneventful recovery and follow up after 3 years, she is doing well.



Figure 3. CXR showing Right Atrium, Right Ventricle enlargement.

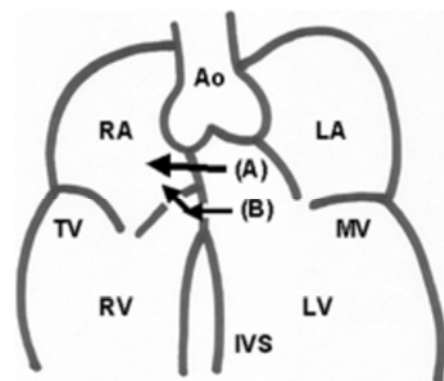


Figure 4. Picture showing types of Gerbode defect. A-Direct type (Supravalvular type) B-Indirect type (Infravalvular type).

4. Discussion

The congenital LV-RA connection was first mentioned in an autopsy report on a patient in 1838 [7, 8]. Riemenschneider and Moss [4] initially classified the defects into two types: Direct and Indirect. Direct defects transcend the membranous septum from the LV to the RA, while indirect defects involve a VSD with accompanying tricuspid regurgitation (TR) [7]. Gerbode defects are later classified by anatomical deficiency of the membranous septum which is divided into two anatomic portions depending on the relationship to the septal leaflet of Tricuspid valve [9-15]. There is more apically located interventricular portion and a more basally placed atrioventricular portion [10, 11, 14]. The tricuspid valve usually attaches to the membranous septum about 1cm apical to the attachment of mitral valve, thus the atrio-ventricular septum separates the LV from RA.

Perimembranous VSD is an obligatory component of two types of communication. The more common type is a defect in the interventricular portion of the membranous septum that opens into RV and then communicates with the RA through the anatomical deficiency in Tricuspid septal leaflet, which is tethered to the crest of the ventricular septum. The defect in Tricuspid valve may be a perforation, a cleft, widened or absence of commissure between Anterior Tricuspid Leaflet and Tricuspid Septal Leaflet. Less common type is direct type of defect in which there is a defect in the atrioventricular portion of membranous septum, so blood from LV enters RA directly [4].

Shunt depends on the size of the defect and PVR. Shunt flow begins in utero because of the obligatory difference in the systolic pressure between LV and RA. RA accepts shunt with less or no elevation of pressures due to distensibility. A high Doppler gradient is one of the hallmarks of the Gerbode ventriculo-atrial defect because of the difference between the left ventricular systolic pressure and the low right atrial pressure. Real-time (RT) three dimensional (3D) echocardiography is more suitable for these anatomical anomalies [16-18]. RT 3D TEE yields rapid, high resolution anatomical characterization of the shunt [16] while providing accurate assessment of the defect's origin, shape, and size; it can also reveal a hidden shunt [18-21].

The defect is associated with slight female preponderance with murmur generated by the shunt is necessarily present at birth. Lesion is susceptible to infective endocarditis. Occasionally, infective endocarditis is responsible for creating Gerbode defect. Raised JVP with RV impulse disproportionately prominent. Split S2 is present on auscultation which widens normally during inspiration. Spontaneous closure of these defects is very rare and surgical repair is excellent.

5. Conclusion

To conclude, Gerbode defect is a rare congenital acyanotic heart disease with a direct communication between LV and RA. A proper echocardiography is needed when RA enlargement is present with a normal LV to diagnose it else it

may be missed for a normal VSD. An early diagnosis and intervention is needed to prevent volume overload and chamber enlargement.

6. Recommendations

Real Time 3D Echocardiography provides accurate assessment of the defect's origin, shape, and size; it can also reveal a hidden shunt.

Consent

Patient gave informed written consent for this publication.

Conflicts of Interest

All the authors do not have any possible conflicts of interest.

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