

Percutaneous Device Closure of Gerbode-type Ventricular Septal Defects

Mariam Opel¹ and Satinder Sandhu²

¹Holtz Children's Hospital

²University of Miami Miller School of Medicine

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Abstract

- The Gerbode type of ventricular septal defect is rare and can be congenital or acquired. - The defect can be closed retrograde or through the transvenous approach. - Short term follow-up suggests that transcatheter closure of the Gerbode type defect is feasible, safe, and effective and should be considered an alternative to surgical repair

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Key Points

- The Gerbode type of ventricular septal defect is rare and can be congenital or acquired.
- The defect can be closed retrograde or through the transvenous approach.
- Short term follow up suggests that transcatheter closure of the Gerbode type defect is feasible, safe and effective and should be considered an alternative to surgical repair

Corresponding Author Information:

Dr. Satinder Sandhu MD

Professor of Pediatrics

Director, Pediatric Cardiac Cath Laboratory

Director, Adult Congenital Heart Disease

1611 NW 12th Avenue, NW Room109

Miami, Florida, 33136

Phone: 305-585-6683

Fax: 305-324-6012

Email: ssandhu3@med.miami.edu

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The Gerbode defect occurs in 0.08% of congenial intracardiac shunts (1) with a higher incidence reported by Yuan et al (2) when acquired defects are taken into account. The septal leaflet of the tricuspid valve attaches to the membranous septum lower than the attachment of the mitral valve. The Gerbode defect results from a deficiency of the atrioventricular (AV) membranous interventricular septum resulting in a left

ventricle to right atrial shunt. The Gerbode defect was initially classified as direct and indirect (3). The direct Gerbode type defect is a deficiency of the atrioventricular (AV) membranous septum. The indirect Gerbode defect is associated with a ventricular septal defect resulting in a perforation of the tricuspid valve from the jet coursing through the ventricular septal defect resulting in a left ventricle to right atrial shunt. This classification was subsequently modified relative to the position of the defect to the tricuspid valve as supra-valvar, infra-valvar or intermediate (4,5). Defects with both supra-valvular and infra-valvular involvement are referred to as intermediate defects. The Gerbode defect should be suspected on transthoracic echocardiogram when there is high doppler velocity from the left ventricle to the right atrium. This defect is unlikely to close spontaneously and can result in volume overload of the right side. It also carries the risk for endocarditis. Successful surgical closure of a left ventricle to right atrial shunt was first described by Kirby et al (6) in 1957 in a patient where the anatomical details were identified intraoperatively. Gerbode et al (7) in 1958 published a case series of 5 patients with a perimembranous ventricular septal defect (pmVSD) that resulted in a left ventricle to right atrial shunt and identified the defect as the “Gerbode defect”. Due to the proximity of the defect to the coronary sinus, conduction system and tricuspid and aortic valves there are anatomical challenges related to transcatheter device closure. In 2006 Trehan et al (8) reported the first transcatheter closure of an acquired Gerbode defect. There are few published case reports and small series reports of immediate and short term results following transcatheter closure of the Gerbode defect.

Haddad et al (9) in a retrospective multicenter study contribute their experience on percutaneous closure of the indirect Gerbode type pmVSD. Transcatheter closure was performed on 10 patients from 3 centers from Aug 2017-May 2021. The patients ranged in age from 2.5-54 years (median age 6.8 years) and in weight from 12-88 Kg (median weight 26.5 Kg). Median left ventricular defect size was 10 mm, (range, 3-15.5 mm). Transthoracic echocardiogram was used to assess the size of defect, presence or absence of the subaortic rim, aortic regurgitation, aortic cusp prolapse, and tricuspid regurgitation. Length deficiency, absence of subaortic rim and tear drop type of aortic valve prolapse with trivial aortic regurgitation was not a contraindication to the device placement. Transcatheter closure of the defect was done under fluoroscopic and TEE guidance and the selection of the device was center and operator based. The devices placed were Amplatzer duct occluder (ADO) II, KONAR-Multifunctional Occluder and ADO I. This is the first report of device closure of the Gerbode defect with the KONAR-Multifunctional occlude. Device placement was retrograde in 9/10 and transvenously through an arteriovenous loop in the single patient with the ADO I device. The diameter of the device placed did not exceed the diameter of the defect on the left ventricular aspect of the septum except in patients with deficiency of the subaortic rim where a device 1-2 mm larger than the defect diameter was placed. At a follow up interval of 3-48 months (median 17 mos) no patient had heart block or tricuspid valve stenosis. There was complete resolution of the shunt and all patients had significant improvement of their tricuspid regurgitation. There was no change in pre-existing aortic regurgitation with one patient developing new onset aortic regurgitation that remained trivial on follow up at 4 years post closure. There were 6 patients with an absent subaortic rim and device placement did not cause aortic regurgitation or interfere with aortic valve function. The authors then go on to comprehensively detail the risks and considerations of closing Gerbode-type defects, listing current literature available while aptly pointing out the lack of homogenized guidelines available when considering this type of pmVSD for transcatheter closure. Haddad et al (9) have effectively demonstrated that percutaneous closure of Gerbode type pmVSD's is a safe and effective alternative to surgical repair and is a valuable addition to the currently available literature available on this type of procedure

Advances in echocardiography have allowed for accurate diagnosis and intra-procedure feedback with regards to optimal device size and device placement to prevent interference with the valves or the conduction system. Vijayalakshmi et al (10) in a series of 12 patients describe successful closure of their pmVSD with one patient developing transient complete heart block that resolved within 48 hours after administration of steroids. The current body of literature does not outline the anatomical characteristics of the defect which would not be amenable to device closure. A wide range of devices have been used for transcatheter closure and the choice of device impacts the procedural approach. The immediate and short-term results of transcatheter closure of the Gerbode defect are reassuring. However, long term follow up is needed.

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