Circulation: Cardiovascular Imaging

CARDIOVASCULAR IMAGES

Tale of 2 Orifices

Three-Dimensional Imaging of Double-Orifice Mitral Valve Types

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Because the congenital double-orifice mitral valve (DOMV) was first reported by William Smith Greenfield in 1876, there have been multiple forms described in the literature.[1] DOMV can be divided into 4 subtypes based on cause: 2 native forms (congenital native and acquired native) and 2 iatrogenic forms (surgical and percutaneous). These subtypes have remarkably similar appearances on 2-dimensional (2D) and 3-dimensional (3D) echocardiography. We describe the historical background and 3D echocardiographic imaging of these various types of DOMV.

CONGENITAL NATIVE DOMV

William Smith Greenfield (1846–1919) was a British pathologist working in London in the late 1800s. Although he was best known for his work on a vaccine for *Bacillus anthracis*, the causative agent of anthrax,[2] he was also the first to describe the congenital DOMV. In an 1876 report, he described a case of a 28-year-old woman who died of myelitis and was found on autopsy to have a curious malformation of the mitral valve. Her mitral valve had 2 orifices instead of 1. He noted that the medial mitral valve orifice was larger than the lateral orifice. He also noted that the valve opened and closed perfectly upon testing with a stream of water.

Although most cases are typically recognized in pediatric age, here we present a case of an incidentally found DOMV in a 72-year-old man with severe symptomatic aortic stenosis because of bicuspid valve seen on transesophageal echocardiography (TEE) imaging before surgical aortic valve replacement. On his 3D TEE, the characteristic orifice size asymmetry of congenital DOMV is well visualized on En face views. In addition, 2D and 3D echocardiography typically demonstrates each orifice with its own tensor apparatus, normal chordae, and papillary muscles. No evidence of significant mitral stenosis was noted on 2D Doppler TEE imaging. (Figure 1, Error! Reference source not found.)

ACQUIRED NATIVE DOMV BECAUSE OF MITRAL VALVE CALCIFICATION

The first description of mitral annular calcification as a pathological entity was by Bonniger[3] in 1908. He described 2 cases of heart block that were thought to be related to deposition of a calcareous hearth within the cardiac conduction system.

When mitral annular calcification predominantly affects the second scallop of the anterior mitral leaflet (A2) and second scallop of the posterior mitral leaflet (P2) portions of the mitral annulus, it can effectively split the mitral valve into 2 orifices. Diastolic restriction of leaflet opening in this region then leads to DOMV.

Key Words: anthrax ■ echocardiography ■ heart block ■ mitral valve ■ papillary muscles

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Here, we present a case of calcific DOMV in a 66-year-old man who had undergone chest radiation for Hodgkin lymphoma and presented with symptomatic mitral stenosis before possible percutaneous valve-in-mitral annular calcification procedure. His calcific DOMV is well seen on both 3D TEE and cardiac computed tomography. Color and spectral Doppler imaging demonstrate significant mitral stenosis. (Figure 2, Error! Reference source not found.)

SURGICAL DOMV BECAUSE OF ALFIERI STITCH

The Italian cardiothoracic surgeon Ottavio Alfieri pioneered the surgical edge-to-edge (e2e) mitral valve repair in 1991 which became to be known as the Alfieri stitch. It is interesting who he came up with idea for this type of repair. It is said that 1 day he had 2 operative cases. The first case was an atrial septal defect closure procedure in a 29-year-old woman, and the second was a complex mitral valve repair of anterior mitral leaflet prolapse. During the first case, Alfieri noted an unexpected finding; in addition to a secundum type atrial septal defect, the patient had congenital DOMV with no significant mitral regurgitation. This provided him with the idea of mimicking this congenital anomaly as a simple method to repair the mitral valve. He subsequently performed e2e mitral valve repair successfully in the second patient by suturing the prolapsed portion of the anterior leaflet to the posterior leaflet. The e2e mitral stitch creates a DOMV.

Here, we present a 68-year-old with severe mitral regurgitation who underwent mitral valve repair with an Alfieri stitch and a 28-mm Calvin-Galloway future band (Medtronic, Minneapolis, MN). His 3D TEE defines the exact anatomic position of a surgical e2e repair and the appearance of a DOMV. Post-e2e mitral repair, there was no significant mitral stenosis. (Figure 3, Error! Reference source not found. and Error! Reference source not found.)

PERCUTANEOUS DOMV BECAUSE OF MITRACLIP

Success of the surgical e2e mitral valve repair technique led to the development of the first percutaneous mitral valve repair system, the MitraClip (Abbott Vascular, Abbott Park, IL). The MitraClip device was invented by the American interventional cardiologist Frederick St Goar, as a percutaneous e2e mitral valve repair in patients deemed too high risk for surgery.

The MitraClip system consists of 3 components: the MitraClip device, a clip delivery system, and a steerable guide. The MitraClip is a polyester fabric-covered, cobalt-chromium device with 2 arms that are opened and closed by a control mechanism. The MitraClip procedure begins with transfemoral venous access, followed by transseptal puncture, and finally clip positioning and implantation using echocardiographic guidance.

Here, we present a case of 74-year-old man with severe symptomatic mitral regurgitation who underwent placement of 2 MitraClips in whom surgical repair was precluded by a high Society of Thoracic Surgery risk score. In him, the 2 clips were placed between the A2 and P2 scallops of the mitral valve, creating a characteristic DOMV on 3D echocardiography. Post-MitraClip, there was no significant mitral stenosis (Figure 4, Error! Reference source not found.).

CONCLUSIONS

In summary, types of DOMV can be divided into 4 categories: congenital native DOMV, acquired native (mitral annular calcification–induced DOMV), surgical (Alfieri stitch e2e repair), and percutaneous (MitraClip e2e repair). Each type has a unique history. Three-dimensional echocardiography is a powerful imaging tool which has the capability of differentiating the various types of DOMV, despite their similar appearances.

ARTICLE INFORMATION

The Data Supplement is available at https://www.ahajournals.org/doi/suppl/10.1161/CIRCIMAGING.118.008372.

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Disclosures

Dr Saric is a member of a Philips and Medtronic speaker's bureau and member of Siemens advisory board. Dr Vainrib is a consultant for Micro Interventional Devices. The other authors report no conflicts.

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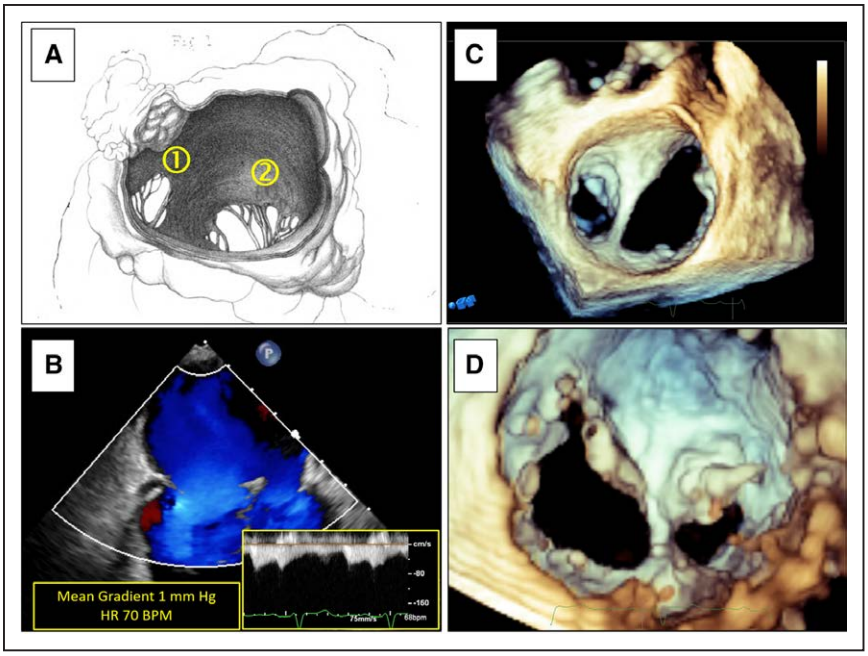

Figure 1. Congenital double-orifice mitral valve (DOMV).
A. William Smith Greenfield, the British physician who provided the original description of the congenital DOMV. **B**. 2D TEE with color Doppler in the midesophageal 60 degree view demonstrates a congenital DOMV. Spectral Doppler demonstrates a normal mean diastolic pressure gradient. **C**. Congenital DOMV from the left atrial side; note the typically unequal orifice size with the lateral orifice being smaller than the medial one. (Video 1 in the Supplementary Digital Content corresponds to this panel.) **D**. Congenital DOMV from the left ventricular side. Video 1 in the Supplementary Digital Content corresponds to this panel. 2D indicates two-dimensional; HR, heart rate; and TEE, transesophageal echocardiogram.

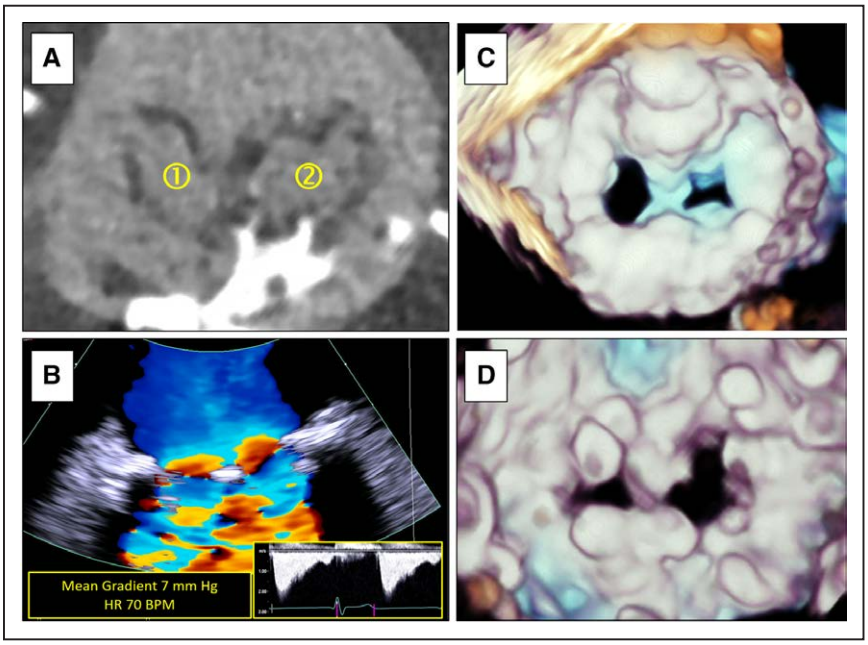

Figure 2. Mitral annular calcification (MAC)–associated double-orifice mitral valve (DOMV).
A. MAC-associated DOMV seen on contrast-enhanced computed tomography of the chest. **B**. 2D TEE with color Doppler in the midesophageal 60 degree view demonstrates a MAC-associated DOMV. Spectral Doppler demonstrates an elevated mean diastolic pressure gradient. **C**. MAC-associated DOMV seen on standard en face surgical view of the mitral valve from the left atrial side. Video 2 in the Supplemental Digital Content corresponds to this panel. **D**. MAC-associated DOMV from the left ventricular side. Video 2 in the Supplemental Digital Content corresponds to this panel. 2D indicates two-dimensional; HR, heart rate; and TEE, transesophageal echocardiogram.

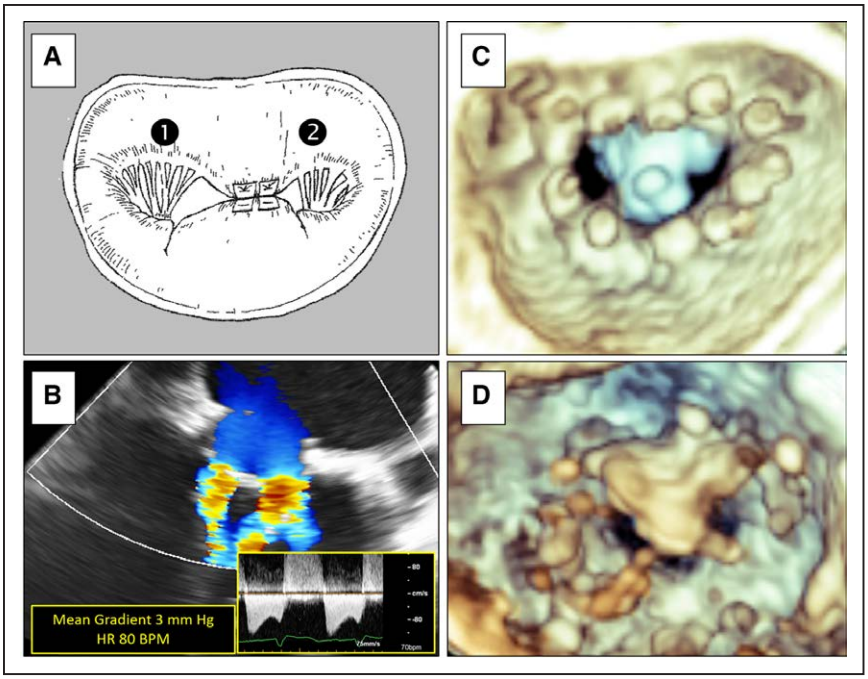

Figure 3. Surgical double-orifice mitral valve (DOMV): Alfieri Stitch.
A. Surgical edge-to-edge repair (Alfieri stitch); the original drawing from the 1995 paper by Alfieri and colleagues. **B**. 2D TEE with color Doppler in the midesophageal 60 degree view demonstrates a DOMV after surgical edge-to-edge repair (Alfieri stitch). Spectral Doppler demonstrates a normal mean diastolic pressure gradient. **C**. Surgical edge-to-edge repair (Alfieri stitch) from the left atrial side. In this patient, the mitral valve repair consisted of a centrally placed Alfieri stich and the CG Future annuloplasty band (Medtronic, Minneapolis, MN). Video 3 in the Supplemental Digital Content corresponds to this panel. Video 4 in the Supplemental Digital Content demonstrates another patient with Alfieri stitch but without annuloplasty band. **D**. Surgical edge-to-edge repair (Alfieri stitch) from the left ventricular side. 2D indicates two-dimensional; HR, heart rate; and TEE, transesophageal echocardiogram.

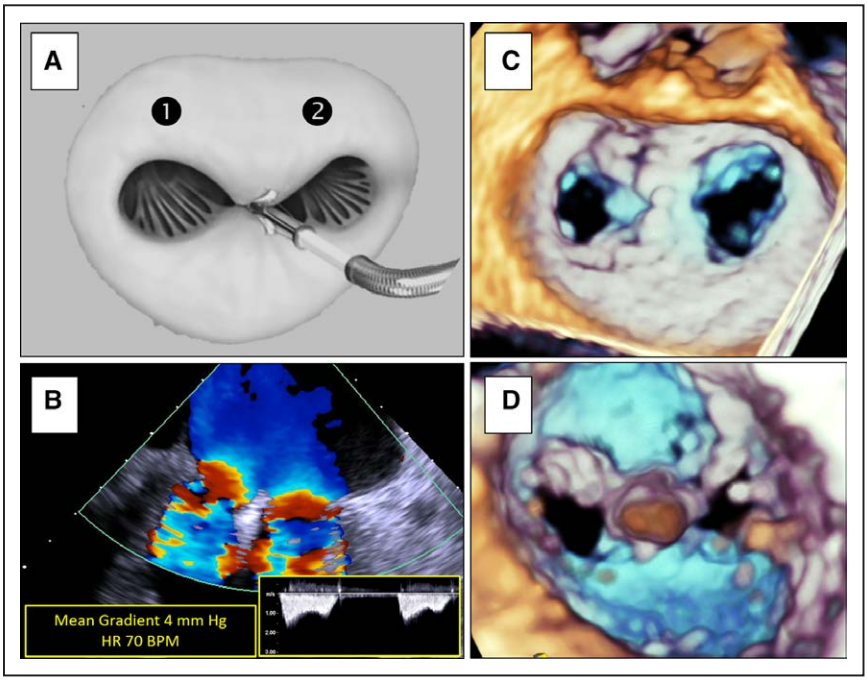

Figure 4. Percutaneous double-orifice mitral valve (DOMV): MitraClip.
A. MitraClip-associated DOMV; drawing courtesy of Abbott Vascular, Santa Clara, CA. **B**. 2D TEE with color Doppler in the midesophageal 60 degree view demonstrates a DOMV after percutaneous edge-to-edge repair (MitraClip). Spectral Doppler demonstrates a normal mean diastolic pressure gradient. **C**. MitraClip-associated DOMV from the left atrial side; the clip is placed centrally across the A2/P2 cooptation line. Video 5 in the Supplemental Digital Content corresponds to this panel. **D**. MitraClip-associated DOMV from the left ventricular side. 2D indicates two-dimensional; HR, heart rate; and TEE, transesophageal echocardiogram.