Acquired Gerbode defect following endocarditis of the tricuspid valve: a case report and literature review

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Abstract
The Gerbode's defect is a communication between the left ventricle and right atrium. It is usually congenital, but rarely is acquired, as a complication of endocarditis, myocardial infarction, trauma, or after previous cardiac surgery. The acquired Gerbode defect with involvement of the tricuspid valve acquired after bacterial endocarditis can be challenging to repair. We present a rare case of young woman, with endocarditis of the tricuspid valve and acquired Gerbode defect without previous cardiac surgery. She underwent successful surgical closure of the Gerbode defect and reconstruction of the septal leaflet of the tricuspid valve using a autologous pericardial patch. A total of 20 other cases were reported with acquired Gerbode defect due to endocarditis in patients without previous cardiac surgery. Three other cases presented acquired Gerbode defect due to myocardial infarction and two due to chest trauma. Another series of 62 patients presented acquired Gerbode defect after previous cardiac surgery. Surgical treatment is always feasible with excellent outcome. However the percutaneous transcatheter closure remains an excellent option especially in high risk patients.

Keywords: Acquired, Gerbode, defect

Introduction
The communication between the left ventricle and right atrium was firstly reported in 1838 by Thurman [1]. In 1957, Gerbode et al. [2] reported the first 5 cases with such a heart defect undergoing successful surgical repair. Such a defect is usually congenital, but rarely is acquired, as a complication of endocarditis [3], myocardial infarction, blunt chest trauma or after previous cardiac surgery [4]. This can be anatomically possible because the normal tricuspid valve is more apically displaced than the mitral valve. Acquired Gerbode defects with large septal destructions and vegetations involving the tricuspid valve can be challenging and might require complex patch repair. We present a case of our patient with this uncommon complication of endocarditis, simulating severe pulmonary hypertension.

Case report
A 40 year old lady from Kosovo, was referred to our hospital for severe pulmonary arterial hypertension and a mass in right atrium suspected for vegetation. About one month before, she was admitted in another hospital and received iv medication. The patient was febrile and the C-reactive protein, white cell count and erythrocyte sedimentation rate were elevated. Blood cultures demonstrated a methacularin sensitive Staphylococcus aureus growth.

Transthoracic echocardiography demonstrated a mobile, irregularly shaped, oscillating and highly mobile mass, located above the tricuspid valve septal leaflet (Fig. 1b). A clear jet across a small defect between left ventricle and right atrium consistent with Gerbode type defect was identified. The direction of the Doppler signal also leads to the true diagnosis (Fig. 1a). Cardiac magnetic resonance demonstrated a supravalvular flow associated with infravalvular jet according to the type C acquired Gerbode defect (Fig. 1c and 1d). A normal lung scan excluded pulmonary embolism. The tricuspid regurgitation was considered mild- to- moderate
with estimated pulmonary arterial systolic pressure about 60-80 mmHg.

The patients underwent surgery after 2 weeks of antibiotic therapy. Through a right atriotomy, large vegetation was attached to the septal leaflet and anterior leaflet of tricuspid valve was identified. On removal of the vegetation, a defect was found communicating between the left ventricle and right atrium (Fig. 2a and 2b). This defect represented an acquired Gerbode defect and was closed by two 5/0 pledgeted prolene sutures (Fig. 2c). Then the septal leaflet of tricuspid valve was resected and was replaced with a trimmed autologous pericardial patch. Anteriorly the newly created septal leaflet was attached to the anterior leaflet. Then, two synthetic chorda were employed (Fig. 2d). The hydraulic maneuver demonstrated trivial tricuspid valve regurgitation (Fig. 2c). Then the right atrium was closed. After an uneventfully post-operative period, the patient was discharged home in good clinical condition. Echocardiogram demonstrated trivial tricuspid valve regurgitation and no residual shunt. One year later the patient was doing well. The transthoracic echocardiography at follow-up demonstrated a moderate tricuspid valve regurgitation and no residual shunt.

**Comment**

Gerbode described such a defect as a congenital atrioventricular shunt originating from the interventricular membranous septum with regurgitation into the right atrium through a defect or cleft in the tricuspid valve leaflet [2]. Less common is the acquired form of a Gerbode defect, which is often associated with bacterial endocarditis [5–24], myocardial infarction [25–27], blunt chest trauma [28, 29] or post previous cardiac surgical procedures [30, 31].

After a careful revision of the literature we found 25 other reported cases with acquired Gerbode defect without previous cardiac surgery. In 22 of them, including our case, the cause was endocarditis. Only 4 patients were females. 7 out 21 cases presented endocarditis due
to *Staphylococcus aureus*, usually involving the aortic valve extending below the aortic annulus onto the upper part of the interventricular septum. Infective tissue destruction leads to a perforation of the septum creating a communication between the left ventricle and the right atrium. However 8 out of 21 cases including our case presented tricuspid valve endocarditis causing an acquired Gerbode defect (Table 1). In difference to the endocarditis of the left side, in the tricuspid valve endocarditis the vegetations and destructed tissue are located in the right side so, it might be more than enough the closure of the communication only on the right side, if healthy tissue is present as in our case. The mortality was almost 9% in patients with endocarditis. Also the postoperative complications such as renal failure was identified in 3 patients (13.6%) and complete atrioventricular block in 3 patients (13.6%). The high incidence of the complete atrioventricular block might be explained with the closed vicinity of the Gerbode defect with the conduction system and atrioventricular node. Interestingly in none of the cases with Gerbode defect without prior cardiac surgery undergoing surgical correction is reported recurrence of the communication between the left ventricle and right atrium or endocarditis recurrence.

Three other cases acquired Gerbode defect post myocardial infarction were found in the literature and all of them presented inferior myocardial infarction. 2 of them died after surgery. Two other patients were found with acquired Gerbode defect due to blunt chest trauma or bullet penetration. The overall mortality in 26 patients without prior cardiac surgery was 15.4%. The postoperative hospital stay was less than 2 weeks in the survived cases.

Interestingly, acquired Gerbode defect after previous cardiac surgery was found in 62 other patients (Table 2). 26 of them underwent surgical closure of the defect and 18 percutaneous closure employing different occlude devices. 11 patients did not undergo any interventional procedure, probably due to small shunt or high operative...
risk. Most of the patients were undergone previously aortic valve surgery or mitral valve surgery. However the mortality, in this group of patients despite all of them were redo operations, was almost 3.2% extremely lower than patients undergoing first time cardiac surgical procedure (Table 1).

The diagnosis was made in most of the cases by transthoracic and transesophageal echocardiography. It seems that echocardiographic examination is the most frequently diagnostic tool employed in these patients. Identification of an actual communication is often extremely difficult, so a careful and meticulous echocardiogram should be done in order to prevent echocardiographic misinterpretation of this defect as pulmonary arterial hypertension. The large systolic pressure gradient between the left ventricle and the right atrium would expectedly result in a high velocity systolic Doppler flow signal in right atrium and it can be sometimes mistakably diagnosed as tricuspid regurgitant jet simulating pulmonary arterial hypertension. However cardiac catheterization, cardiac tomography or magnetic resonance such as in our case offers valuable information. Interestingly our case after been diagnosed with Gerbode defect underwent cardiac magnetic resonance which revealed a class C acquired Gerbode defect as previously described [4].

Treatment of the acquired Gerbode defect depends on symptoms, magnitude of shunt, flow volume, concomitant anatomic abnormalities and co-morbidities. Asymptomatic, chronic, small defects can be managed conservatively.

Percutaneous transcatheter closure techniques have been employed in almost 25% of patients, mostly in high risk surgical candidates due to previous valve replacement, advanced age, anti-coagulation, and multiple co-morbidities. Advanced cardiac imaging techniques such

### Table 1 Patients with acquired Gerbode defect without prior cardiac surgery

| Author (Ref) | Year | Gender/Age | Location | Bacteria | Diagnosis | Treatment | Outcome |
|-------------|------|------------|----------|----------|-----------|-----------|---------|
| Battin [5]  | 1991 | Male/15    | na       | na       | TTE       | Surgery   | Survived |
| Saiki [6]   | 1994 | Male/42    | MV/AV    | Streptococcus hemolyticus | TTE, Surgery | Survived |
| Katoh [7]   | 1994 | Male/58    | TV       | na       | TTE       | Surgery   | Survived |
| Elian [8]   | 1995 | Male/64    | TV       | Staphylococcus aureus    | TTE, TEE, CC | Surgery | Survived |
| Velebit [9] | 1995 | Male/30    | BAV      | Staphylococcus aureus | TEE, CC | Surgery | Survived(AVB) |
| Winslow [10]| 1995 | Male/30    | AV       | Staphylococcus aureus | TTE, TEE | Survivor |
| Michel [11] | 1996 | Male/52    | AV       | Streptococcus viridans | TTE, TEE | Conservative | Survived |
| Alphonso [12]| 2003 | Male/63    | AV       | Culture negative | TTE | Surgery | Survived |
| Raja [13]   | 2006 | Male/47    | RA       | Staphylococcus aureus | TTE, TEE | Survivor | Survived(RF) |
| Fukui [14]  | 2007 | Male/57    | TV, AV, MV | na       | TEE       | Surgery | Survived |
| Tatewaki [15]| 2008 | Female/7   | TV, AV, MV | Staphylococcus aureus | TTE, CT | Surgery | Survived |
| Inouel [16] | 2009 | Female/21  | AV       | Culture negative | TTE, TEE | Survivor | Survived |
| Cortez-Dias [17]| 2009 | Male/59   | MV       | Staphylococcus aureus | TTE, TEE | Conservative | Died(AVB, RF) |
| Mendoza [18]| 2009 | Female/52  | AV       | Streptococcus mutans | TTE, CT | Surgery | Survived |
| Hori [19]   | 2010 | Male/41    | BAV      | na       | TTE       | Surgery   | Survived |
| Matt [20]   | 2010 | Male/35    | AV       | Hemophilus aphrophilus | TTE,TEE | Survivor | Survived(AVB) |
| Ota [21]    | 2011 | Male/71    | AV       | Streptococcus pneumonia | TTE,TEE | Surgery | Survived |
| Pillai [22] | 2011 | Male/12    | TV       | Culture negative | TEE | Surgery | Survived |
| Carpenter [23]| 2012 | Male/22   | TV       | Staphylococcus lugdunensis | TEE, CT | Survivor |
| Hsu [24]    | 2014 | Male/40    | BAV      | Cardiobacterium hominis | TEE | Surgery | Died(RF) |
| Prifti et al. | 2015 | Female/40  | TV       | Staphylococcus aureus | TTE, TEE | Survivor |

### Area of myocardial infarction

| Author (Ref) | Year | Gender/Age | Location | Bacteria | Diagnosis | Treatment | Outcome |
|-------------|------|------------|----------|----------|-----------|-----------|---------|
| Hole [25]   | 1995 | Male/63    | Inferior myocardial infarction | na       | TTE       | Surgery   | Survived |
| Jobic [26]  | 1997 | Female/72  | Inferior myocardial infarction | na       | TTE, TEE | Surgery | Died (RF) |
| Newman [27] | 1996 | Male/72    | Inferior myocardial infarction Trauma | na       | TTE, TEE | Surgery | Died |
| Venkatesh [28]| 1996 | Male/16    | Blunt trauma | na       | TTE, TEE | Surgery | Survived |
| Selinger [29]| 1998 | Male/70    | Bullet, trauma | na       | TTE,TEE,CC | Surgery | Survived |

Legend: TTE Transthoracic echocardiography, TEE Transesophageal echocardiography, CC Cardiac catheterization, CT Cardiac tomography, na not available, AV Aortic valve, BAV Bicuspid Aortic Valve, MV Mitral valve, TV Tricuspid valve, RF Renal Failure, AVB Complete atrioventricular block
| Author                  | Year | Gender | Age | Diagnostic tool | Previous procedure | Treatment       | Outcome   |
|------------------------|------|--------|-----|-----------------|--------------------|-----------------|-----------|
| 1. Katta et al.        | 1994 | Male   | 54  | TTE, TEE        | Endomyocardial biopsy | Conservative    | Survived  |
| 2. Dzwonczyk et al.    | 1995 | Male   | 25  | TTE             | ASD repair         | na              | na        |
| 3. Dzwonczyk et al.    | 1995 | Female | 72  | TTE             | AVR, VSD repair    | na              | na        |
| 4. Fukui et al.        | 2000 | Male   | 53  | TEE             | MVR x 2             | Surgery         | Survived  |
| 5. Benisty et al.      | 2000 | Male   | 72  | TEE, TEE        | MVR                | Surgery         | n.a.      |
| 6. Benisty et al.      | 2000 | Male   | 73  | TEE, TEE        | MVR x 3, AVR       | Surgery         | n.a.      |
| 7. Weinrich et al.     | 2001 | Female | 58  | TEE, CC         | MVRx 2              | Surgery         | Survived  |
| 8. Wasserman et al.    | 2002 | Male   | 78  | TTE, TEE, AVR   |                    | Surgery         | Survived  |
| 9. Cabalka et al.      | 2005 | Female | 70  | TEE, TEE        | MVR x 2             | Percutaneous    | Survived  |
| 10. Lorber et al.      | 2006 | Female | 78  | TEE, CC         | MVR                | Percutaneous    | Survived  |
| 11. Ramasubbu et al.   | 2006 | Male   | 41  | TEE             | Aortic root        | Surgery         | Survived  |
| 12. Ramasubbu et al.   | 2006 | Female | 51  | TEE             | Aortic root        | Conservative    | Survived  |
| 13. Trehan et al.      | 2006 | Male   | 22  | TTE, MRI, CC    | VSD + sinus valsalva repair | Percutaneous | Survived  |
| 14. Martinez et al.    | 2007 | Female | 70  | TTE             | MVR                | Percutaneous    | Survived  |
| 15. Martinez et al.    | 2007 | Male   | 67  | TEE             | MVR                | Percutaneous    | Survived  |
| 16. Uslu et al.        | 2007 | Male   | 54  | TEE             | MVR                | Surgery         | Survived  |
| 17. Hilberath et al.   | 2007 | Male   | 68  | TEE             | AVR + endocarditis | Surgery         | Survived  |
| 18. Frigg et al.       | 2008 | Female | 77  | TEE, CC         | AVR                | Surgery         | Survived  |
| 19. Moaref et al.      | 2008 | Female | 51  | TEE             | MVR                | Surgery         | n.a.      |
| 20. Aoyagi et al.      | 2008 | Female | 71  | TTE, CC         | MVR, TV repair      | Surgery         | Survived  |
| 21. Rothman et al.     | 2008 | Male   | 86  | TTE, CC         | MVR                | Percutaneous    | Survived  |
| 22. Hansalia et al.    | 2009 | Female | 46  | TEE             | AVR                | Surgery         | Survived  |
| 23. Yared et al.       | 2009 | Male   | 60  | TTE, TTE        | AVRx + endocarditis | na             | na        |
| 24. Gorhri et al.      | 2009 | Female | 69  | na              | AVRx + endocarditis | na             | na        |
| 25. Subramaniam et al. | 2009 | Male   | 60  | TEE, CT         | AVR                | Surgery         | Survived  |
| 26. Amirghofran et al. | 2009 | Female | 51  | TEE             | MVR                | Surgery         | Survived  |
| 27. Silbiger et al.    | 2009 | Female | 30  | TTE, CC         | VSD repair         | Conservative    | Survived  |
| 28. Cheema et al.      | 2009 | Female | 31  | MRI             | VSD repair         | Conservative    | Survived  |
| 29. Can et al.         | 2009 | Male   | 72  | TTE             | AV nod ablation    | Conservative    | Survived  |
| 30. Can et al.         | 2009 | Male   | 68  | Autopsy         | AV nod ablation    | na             | Died      |
| 31. Dadkhah et al.     | 2009 | Female | 73  | TEE             | TV repair          | Conservative    | Survived  |
| 32. Mohapatra et al    | 2009 | Female | 22  | TEE             | MVR (RF)           | Surgery         | Survived  |
| 33. Sun et al.         | 2010 | na     | na  | na              | MVR                | Surgery         | na        |
| 34. Sun et al.         | 2010 | na     | na  | MVR             | na                 | na             | na        |
| 35. Pursnani et al.    | 2010 | Male   | 78  | TTE, TEE        | AVR                | Surgery         | Survived  |
| 36. Sharma et al.      | 2011 | Male   | 80  | TTE             | AV nod ablation    | Conservative    | Survived  |
| 37. Kumar et al.       | 2011 | Female | 59  | TEE             | AVRx2 + endocarditis | Surgery    | Survived  |
| 38. Zhu et al.         | 2012 | Baby   | 6 months | TTE, TEE | ASD, VSD repair    | Percutaneous    | Survived  |
| 39. Bochard-Villaneuva | 2012 | Male   | 63  | TEE, CT         | AVRx + endocarditis | Surgery    | Survived  |
| 40. Vallakati et al.   | 2012 | Female | 53  | TTE             | AVR                | Conservative    | Survived  |
| 41. Elmistekawy et al. | 2012 | Male   | 59  | TEE             | AVR                | Surgery         | Survived  |
| 42. Dore et al.        | 2012 | Male   | 50  | TTE, TEE        | AVR, MVR           | Surgery         | Survived  |
| 43. Yurdakul et al.    | 2012 | Male   | 68  | TEE             | AVR                | Surgery         | Survived  |
| 44. Mousavi et al.     | 2012 | Female | 76  | TEE, MRI        | AVR                | Conservative    | Survived  |
as transesophageal echocardiography provide excellent images for guidance in device sizing and deployment. The Amplatzer duct occluder device is a mainstay in treatment as it provides less radial force [30] than the muscular ventricular septal defect closure device causing fewer complications [31].

In most of the cases with acquired Gerbode defect a simple direct suture might be enough to close the defect [12] such as in our case, although large Gerbode defect associated with partial or total destruction of the tricuspid valve can be much more challenging. In such cases reconstruction or replacement of the tricuspid valve might be required. Tatewaki et al. [15] describe a pericardial patch closure with sutures from the ventricular side of the tricuspid valve through the leaflets. Others reported a Dacron patch closure with septal leaflet reimplantation onto the patch [9, 12], an annuloplasty ring implantation, or tricuspid valve replacement [5, 8, 9, 12]. Matt et al. [20] presented a double plicated patch combining a defect closure and reconstruction of the tricuspid valve annulus and septal leaflet. In our case we closed the defect from the right side using two single pledged prolene suture and reconstruct the septal and anterior tricuspid valve leaflets using an autologous pericardial patch. This technique allowed us to perform a complex right-sided defect repair with one patch that might be advantageous in an infective situation. Such a technique might allow an extensive reconstruction of the tricuspid valve, if necessary.

As conclusion, the acquired Gerbode defect a rare form of intracardiac shunt, but its incidence has been increasing during the last decades. Increased numbers of invasive and repeat cardiovascular procedures and infective endocarditis have led to this increase in acquired Gerbode defect. Surgical treatment is always feasible with excellent outcome. However the percutaneous transcatheter closure remains an excellent option especially in high risk patients.

**Conclusion**

The acquired Gerbode defect a rare form of intracardiac shunt, but its incidence has been increasing during the last decades. Increased numbers of invasive and repeat cardiovascular procedures and infective endocarditis have led to this increase in acquired Gerbode defect. Surgical treatment is always feasible with excellent outcome. However the percutaneous transcatheter closure remains an excellent option especially in high risk patients.

**Consent**

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of the Journal of Cardiothoracic Surgery.

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**Table 2 Patients with acquired Gerbode defect undergoing previous cardiac surgery (Continued)**

| 45. Ozdogan et al. | 2012 | Female | 31 | TTE, TEE | MVRx2 + endocarditis | Surgery | Died |
|-------------------|------|--------|----|----------|----------------------|---------|------|
| 46. Anderson et al. | 2012 | na     | na | na       | AVR                  | na      | na   |
| 47. Toprak et al. | 2013 | Male   | 32 | TTE, TEE | AVR                  | Conservative | Survived |
| 48. Notarangelo et al. | 2013 | na     | 69 | TTE, TEE | MVR                  | Percutaneous | Survived |
| 49. Sinisalo et al. | 2013 | Male   | 75 | TTE, TEE, CC | AVR                  | Percutaneous | Survived |
| 50. Sinisalo et al. | 2013 | Female | 23 | TEE, CC | VSD repair         | Percutaneous | Survived |
| 51. Sinisalo et al. | 2013 | Male   | 10 | TEE, CC | ASD, VSD repair     | Percutaneous | Survived |
| 52. Sinisalo et al. | 2013 | Male   | 8  | TEE, CC | VSD repair         | Percutaneous | Survived |
| 53. Dangel et al. | 2013 | Male   | 6 months | TTE, TEE, CC | ToF repair     | Percutaneous | Survived |
| 54. Lee et al. | 2013 | Male   | 3 months | TTE, CC | ASD, PDA, VSD repair | Percutaneous | Survived |
| 55. Poulin et al. | 2013 | Female | 75 | TTE, TEE | MVR                  | Percutaneous | Survived |
| 56. Primus et al. | 2013 | Female | 76 | TTE, TEE | AVR                  | Conservative | Survived |
| 57. Chaturvedi et al. | 2013 | Male   | 62 | TTE, MRI | AVR                  | Percutaneous | Survived |
| 58. Tayama et al. | 2014 | Male   | 75 | TTE, CC | MV and TV repair    | Surgery | Survived |
| 59. Hussein et al. | 2014 | Male   | 45 | TTE, TEE | AVRx2                | Surgery | Survived |
| 60. Chamsi-Pasha et al | 2014 | Male   | 67 | TTE, TEE | MVR, TVR             | Surgery | Survived |
| 61. Taskesen et al. | 2014 | Male   | 74 | TTE, TEE | AVRx2                | Percutaneous | Survived |
| 62. Fanari et al | 2015 | Female | 50 | TTE, CT | AVR                  | Percutaneous | Survived |

Patients with acquired Gerbode defect undergoing previous cardiac surgery

Legend: TTE Transthoracic echocardiography, TEE Transesophageal echocardiography, CC Cardiac catheterization, CT Cardiac tomography, MRI Magnetic resonance, na-not available, AVR Aortic valve replacement, MVR Mitral valve replacement, TV Tricuspid valve, ASD Atrial septal defect, VSD Ventricular septal defect, ToF Tetralogy of Fallot, PDA Patent ductus arteriosum
Competing interests
We declare that we do not have any competing or financial interests with this manuscript.

Authors’ contribution
FA and AD diagnosed the patient and followed the patient postoperatively. EP and AB performed the surgery. All authors were involved on the literature review and manuscript writing process. All authors read and approved the final manuscript.

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