Current Cardiology Reports (2018) 20: 90
https://doi.org/10.1007/s11886-018-1038-z

STRUCTURAL HEART DISEASE (RJ SIEGEL AND NC WUNDERLICH, SECTION EDITORS)

Ruptured Sinus of Valsalva Aneurysm and Gerbode Defects: Patient and Procedural Selection: the Key to Optimising Outcomes

Colm R. Breatnach[1] · Kevin P. Walsh[1]

Published online: 20 August 2018

Abstract

Purpose of Review In this review, we reflect on the historical background, clinical features and imaging techniques used to assess Gerbode defects and sinus of Valsalva aneurysms. We aim to review the evolution of treatment strategies and the progression towards less invasive management for these conditions.

Recent Findings While transthoracic echocardiography is often diagnostic, transesophageal echocardiography (2D and 3D) has improved our understanding of these defects and allowed us to more accurately define their anatomy. Cardiac MRI provides improved assessment of the physiological impact of defects by quantifying shunt volume. Transcatheter techniques are currently vying with surgery as the mainstay of treatment.

Summary New insights are being discovered regarding diagnostic modalities and treatment pathways. Defining criteria for patient selection for catheter or surgical therapy is essential when deciding on the optimum intervention for the individual patient.

Keywords Sinus of valsalva aneurysm · Gerbode defect · Review article · Treatment strategies

Abbreviations

ADO	Amplatzer duct occluder
AR	aortic regurgitation
ASD	atrial septal defect
CCF	congestive cardiac failure
LA	left atrium
LV	left ventricle
PC	percutaneous closure
PAH	pulmonary artery hypertension
RA	right atrium
SOVA	sinus of valsalva aneurysm
TEE	transesophageal echocardiography
TTE	transthoracic echocardiography
TR	tricuspid regurgitation
VSD	ventricular septal defect

This article is part of the Topical Collection on *Structural Heart Disease*

✉ Kevin P. Walsh
kpjwalsh@me.com

Colm R. Breatnach
colm.breatnach@gmail.com

[1] Our Lady's Children's Hospital, Dublin, Ireland

Gerbode Defect

Introduction

The discovery of a left ventricular (LV) to right atrial (RA) communication was first described at autopsy by Thurman et al. in 1838. Very little additional information was discovered about this defect until a further autopsy series in five patients by Perry et al. This led to the classification of the lesion in 1949 into four subtypes [1]. Frank Gerbode documented the first series describing surgical repair of the defect in 1958, and the lesion was subsequently named the "Gerbode defect" [2]. Sakakibara and Kono later refined the classification of the lesion into three categories. They described a supravalvar defect (type 1) with a deficiency in the atrioventricular septum above the septal leaflet of the tricuspid valve in the region where off setting of the atrioventricular valves occurs (Fig. 1); an infravalvar defect (type 2), in which there is a perimembranous ventricular septal defect (VSD) with an additional deficiency in the tricuspid valve leaflet, and a combined type of defect (type 3) [3]. Type 1 lesions allow for direct shunting of blood from the LV to RA. In type 2 lesions, the shunted blood crossing the VSD is directed through the deficiency in the tricuspid valve septal leaflet into the RA. Although type 2 (indirect) lesions were previously believed

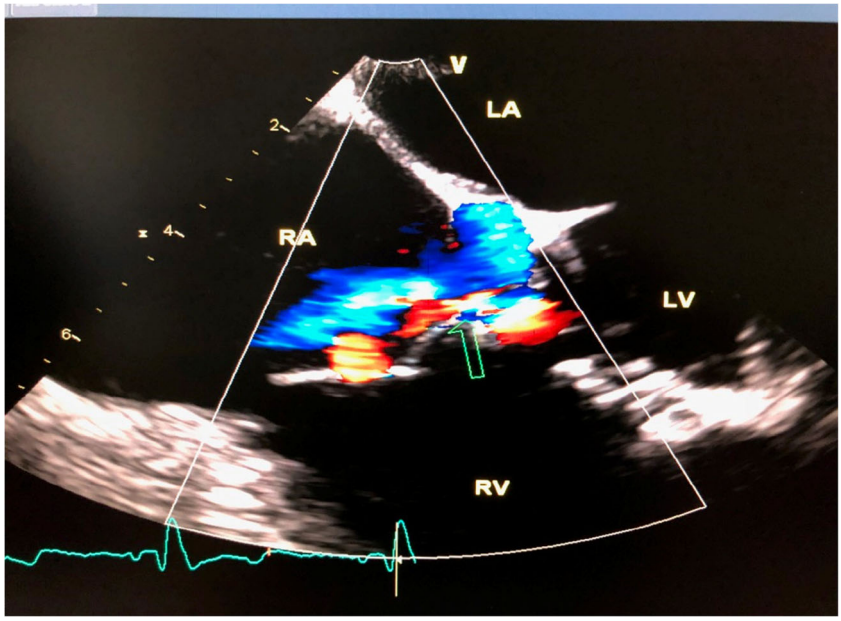

Fig. 1 Transesophageal image demonstrating a direct (type I) Gerbode defect. The *red jet* reveals the left ventricular to right atrial shunt

to be more frequent than type 1 (direct), recent reports on the frequency of these defects state an incidence of 76% direct, 16% indirect and 8% combined [4].

Congenital Gerbode Defects

Congenital Gerbode defects are responsible for less than 1% of all congenital heart lesions [5]. They may be either direct or indirect LV to RA communications [6]. It is recognised that the congenital variety of Gerbode defect is complex and multifactorial [7]. Genetics may play a role in the developmental process. A genotype-phenotype correlation in patients with a Gerbode VSD was investigated by Borkar et al. They performed molecular studies which demonstrated that the "contributing sequence variations in the NKX2-5, GAGA4 and TBX5 gene lies in one of the highly conserved regions and this region is responsible for encoding a functional protein" [8]. Their team concluded that the resulting genotype variation might be linked with the formation of the Gerbode defect. Regression of smaller defects has been known to occur due to tricuspid valve tissue hyperplasia and aneurysm formation [9].

Acquired Gerbode Defects

The incidence of acquired defects is less well defined [4]. As with congenital lesions, acquired Gerbode defects may also be direct or indirect. A comprehensive literature review describing acquired Gerbode defects in 2015 performed by Yuan described 237 cases with a male to female ratio of 2.2:1. Acquired LV-RA shunts may result from a variety of causes including: iatrogenic 51%, infective 37%, traumatic 9% and ischemic 3%. In Yuan et al. review, 81% were direct, 12% indirect and 7% were combined [10••]. The shunt developed at 20.5 ± 38.1 months (median 2; range 0–408) ($n = 49$) after the impact of the inducing factor.

Common surgical causes of Gerbode defects include: VSD closure, mitral and aortic valve procedures [11]. Other reported procedural causes include myomectomy and coronary artery bypass grafting. The review by Yuan also highlighted cardiac catheterisation as a potential cause of Gerbode defects. Radiofrequency catheter ablation of the atrioventricular node was noted in four patients and endomyocardial biopsy after heart transplantation in one patient [12–14]. Infective endocarditis (IE) is another significant contributor to the development of these communications. The latency period for the formation of an infective LV-RA shunt can be very short [10••]. IE must always be considered in newly diagnosed lesions.

Clinical Presentation

Although the anatomy of direct and indirect defects is unique and important to define on imaging, the physiological implications are very similar. With direct communications, blood shunts directly from the LV to the RA. In indirect defects, blood shunts from the LV across the VSD and into the RA via a deficient tricuspid valve septal leaflet. The large pressure gradient can result in a large volume of shunted blood into the right heart [15].

Symptoms vary dramatically depending on the size of the defect and volume of additional pulmonary blood flow (Qp:Qs) [4]. Small defects may be asymptomatic [16], but most patients will present with symptoms of pulmonary over-circulation including: dyspnea, fatigue and weakness, with chest pain also described. Fever is a relatively frequent

finding and care must be taken to out rule IE [4, 10]. The typical examination finding is of a harsh pansystolic murmur along the left sternal border. This is demonstrated in 72% of patients [10••, 17]. Features of elevated RA pressures include elevated jugular venous pulsations, hepatomegally and peripheral oedema [18].

Investigations

Transthoracic echocardiography (TTE) is frequently performed as a first-line imaging modality for Gerbode defects. However, caution must be exercised when performing echocardiography in these patients. The high velocity LV-RA jet may be misdiagnosed as tricuspid regurgitation (TR) with pulmonary artery hypertension (PAH) [19]. Mahajan et al. noted that if two separate TR jets with different timings in systole are present, this may help in "differentiating an infravalvular (indirect) type of Gerbode defect from an Eisenmenger VSD with TR" [20].

The physiology of LV-RA shunts will typically result in RA dilation clearly visible on TTE [5]. If large shunts are present, i.e. those with a Qp:Qs > 2:1, over-circulation of blood to the lungs and increase venous return to the left atrium (LA) may also lead to left heart volume overload. A significant left to right shunt may precipitate PAH over time [21]. Several additional lesions must be assessed when imaging patients with a Gerbode defect including: valvular leaflet perforation and subannular abscess [5].

Transesophageal echocardiography (TEE) (Fig. 1) has displayed a greater precision than transthoracic imaging. A large literature review demonstrated that TEE was the most sensitive diagnostic technique for assessing Gerbode defects, with "less diagnostic bias in the diagnosis of this lesion" [5]. In a review of imaging modalities utilised in Gerbode defects, Taskesen et al. highlight the merit of 3D TEE to visualise the defect in the "en face" view. They describe it as the main modality in diagnosis and decision making for this defect and identify its role in transcatheter closure of the defect [16].

Further definition of shunt anatomy, quantification of shunt ratio and accurate measurement of ventricular volumes may be ascertained with the use of cardiac magnetic resonance imaging which is frequently performed [22]. It is, however, more costly and time consuming, with limited availability in some centres [16].

Management

Because of the rarity of Gerbode defects, much of what we know regarding treatment is based on case reports or series. Yacoub et al. proposed that all LV-RA defects should be repaired, regardless of their size, to prevent the risk of IE and due to the low probability of spontaneous closure. In addition, surgical repair is considered relatively safe with a very low morbidity and mortality [23]. Toprak et al., however, proposed that patients with an insignificant LV-RA shunt, who were asymtopmatic, with no echocardiographic features of right ventricular volume or pressure loading did not mandate surgical closure and could be closely followed [24]. This is somewhat supported by the evidence of spontaneous closure of Gerbode defects by surgical exploration [25]. The closure of congenital defects may occur due to tricuspid septal leaflet aneurysm transformation walling off the defect; however, the incidence of IE is high [26]. It is important to note that spontaneous closures has been reported to be less likely in type I LV-RA defects and more likely in acquired than in congenital shunts [27].

Surgical Closure

Surgical repair of Gerbode defects has demonstrated excellent outcomes [7]. When surgical closure is performed, smaller defects may be closed by suture repair alone, with patch repair utilised for moderate to large defects. It is recommended that the patch be sutured on to the right atrial side of the defect in order to prevent postoperative heart block and to reduce the risk of recurrence [7, 10]. Several different surgical approaches have been described including: Dacron patch closure with septal leaflet re-implantation; annuloplasty ring implantation or tricuspid valve replacement [7, 28]. Of note, in patients with infective endocarditis, paravalvular extension is associated with a high mortality if managed medically without surgical intervention [29]. As a result, surgical management of all acquired Gerbode defects secondary to IE is advised [30].

Percutaneous Closure

The first device closure of an acquired LV-RA communication was reported in 2006 [21]. Since then, percutaneous closure (PC) of Gerbode defects has become a more frequent procedure, avoiding the need for sternotomy and bypass. A variety of devices have been employed depending on defect size, proximity to conduction tissue and operator preference. These devices include the: Amplatzer ductal occluder (ADO) I, ADO II, Amplatzer muscular VSD occluder device, Cera ductal occluder and Nit-occlud Le VSD coil [4, 30–34]. It has been suggested that the ADO may be more suitable than the muscular VSD device because of its low radial force [34]. In addition, Vijayalakshmi et al. strongly advocate for the ADO II device when performing PC due to its low profile and soft retention discs. They report a series of 12 successful cases of closure in patients from 10 months to 16 years of age using a retrograde approach [35•]. Device closure is not limited to the adult or adolescent population and has recently been reported in a 3-month-old infant weighing 3 kg using an ADO [6]. In a review of 234 patients by Yuan, PC of Gerbode defects were only performed in patients with iatrogenic defects. Surgical treatment was the main approach in those with IE. Forty patients

did not receive an intervention. Yuan noted that the PC group had higher rates of complication, mainly relating to a higher incidence of residual shunts. Post-operative heart block was higher in the surgically treated group. Mortality was highest in those receiving no intervention [10••].

Conclusion

Although a rare disease, Gerbode defects have important clinical implications with a high mortality rate if left untreated. Understanding the mechanism of disease is important and care must be taken to determine the presence of IE. Several imaging modalities are available to help define the precise anatomy of the defect, essential when considering appropriate intervention. Surgical closure is well described with a high success rate and should be pursued for complex lesions or those with IE. The development of new devices has allowed for safe and successful device closure in select patient groups.

Sinus of Valsalva Aneurysm

Introduction

Sinus of Valsalva aneurysms (SOVA) are a congenital cardiac defect that arise due to a lack of continuity between the aortic media and annulus fibrosa secondary to incomplete fusion. They may also form due to a structural defect within the aortic annulus. SOVA was first reported in the mid-nineteenth century. In 1840, Thurnam described the clinical features of the condition and noted the high prevalence of aneurysm formation within the right and non-coronary cusps [36]. In 1957, Edwards and Burchell provided further morphological data on the condition in their work on autopsy specimens [37]. They believed that the lack of continuity between the aortic media and annulus fibrosa predisposed these patients to the development of SOVAs. Progression to rupture at this weakened site may evolve if left unrepaired. The first surgical repair was described by Lillehei et al. in 1957 [38]. Traditionally, surgical repair has been the mainstay of treatment although since the first PC in 1994, a transcatheter approach has become a viable option for certain cases [39].

Background of Sinus of Valsalva Aneurysms

SOVA are present in approximately 0.15–1.5% of patients undergoing bypass. They are more often congenital than acquired in origin. Acquired causes include infective endocarditis (IE), syphilis, trauma, atherosclerosis, cystic medial necrosis and connective tissue disorders including Marfans syndrome. In one study, Marfans syndrome was present in 9% of patients [40]. The commonest acquired cause reported in the literature is IE [40, 41]. There is a male preponderance, with a male to female ratio between 2:1 and 3:1 [40, 42]. The incidence of SOVA is significantly higher among the Asian population with a 5-fold increase in reported cases than western populations. The onset of rupture also appears to be earlier in Asian patients with a reported age of rupture between 27 and 32 years in Japanese and Chinese patients, respectively, compared with 42 years in American patients [43, 44]. Takach et al. reviewed 129 patients over a 40-year period and noted a rupture rate of 49.6% in their cohort [40]. Moustafa et al. reported their experience of SOVA in the Mayo clinic. Within their patient cohort, they identified 70% arising from the right coronary sinus, 26% from the non-coronary sinus and 5% from the left coronary sinus.

Clinical Presentation

The clinical presentation of patients with this defect is varied. Prior to rupture, patients will frequently demonstrate no clinical manifestations. When symptoms do arise, it is typically due to compression or rupture of the aneurysm. This most commonly occurs in young adult life but has been reported at any age [45]. Once rupture of the aneurysm has occurred, the onset of symptoms may be abrupt with nearly 80% of patients demonstrating symptoms. [46]. As the typical location of rupture occurs in the right or non-coronary sinus (into the RA or RV), the classic physiology is of a left to right shunt [40]. This may have profound hemodynamic effects, resulting in symptoms of congestive cardiac failure (CCF), including exertional dyspnea and fatigue. Palpitations and chest pain are also described [45, 47]. Clinical examination may reveal the presence of a continuous murmur.

If left untreated, progression to heart failure and sudden death in nearly 80% may occur [31]. Death from CCF will often occur within 1 year of rupture, although longer survival times have been described in those with slow perforation. Less typical presenting features of SOVA are due to complications of the defect and include acute coronary syndrome, cardiac tamponade, aortic regurgitation (AR), heart block and right ventricular outflow tract obstruction [48]. Case reports in the literature have also described cerebral and coronary thrombus embolization caused by unruptured aneurysms [49, 50]. The risk of thrombus is believed to be due to the "abnormal vortex flow in the sinus of Valsalva aneurysm" [51].

Investigations

Transthoracic echocardiography (TTE) is diagnostic, although TEE is a helpful adjunct to define the anatomy of the defect (Fig. 2). Prior to rupture, the classic "windsock" appearance is seen. Following rupture, left to right shunting across the defect is visualised. Echocardiography may also unveil associated lesions, including atrial septal defects (ASDs), bicuspid aortic valves in 16%, VSDs in 12% and aortic root dilation [40, 45,

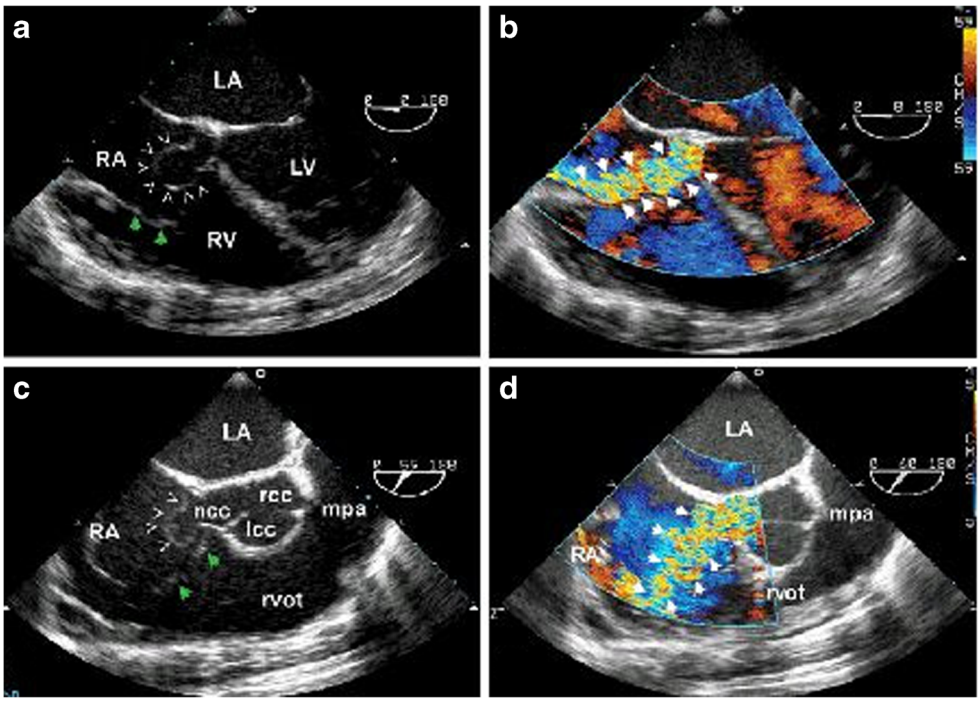

Fig. 2 Transesophageal echocardiogram demonstrating a ruptured sinus of Valsalva aneurysm in 2D and colour Doppler imaging. **a**, **c** 2D images of a classic windsock appearance of a SOVA protruding into the right atrium in the 4 chamber (**a**) and short axis (**c**) views, respectively. **b**, **d** Colour Doppler images in the same views demonstrating rupture of the aneurysm with high velocity shunting from the aorta into the right atrium

47, 52, 53]. In a review by Moustafa et al., 65% of patients were diagnosed by echocardiography and the remainder by cardiac catheterisation [45]. Studies have demonstrated that in patients with SOVA of Chinese and Indian ethnicity, an associated VSD was almost always subarterial, whereas perimembranous defects predominate in western patients [41].

Management

Historically, surgical repair was advocated in all patients when a diagnosis of SOVA was found [40, 45]. Other groups have discussed a more conservative approach, favouring repair only in patients with symptoms, compression of adjacent structures or when the aneurysm is more than 50% of the average size of the other sinuses. These groups suggest that those with unruptured, stable aneurysms should receive anticoagulation and 6-month review [54].

Surgical Repair

Surgical options include direct suture approximation or the integration of patches (pericardial or prosthetic). The risk of recurrence with either approach is debated [43, 47, 52, 53, 55]. Mortality rates have been low with 25-year survival reported at 86% [52], with a further improvement in more recent survival figures (43). The survival rate at 10 years after surgery could now reach up to 90% [56, 57]. In a retrospective review assessing long-term outcomes in patients with surgically corrected SOVA, Moustafa et al. noted a median age of repair of 45 years with pre-operative rupture identified in 34% of the cohort. Eighty-four percent underwent an additional cardiac procedure during the initial repair. The majority of aneurysm rupture occurred into the RV [45]. Additional procedures at the time of aneurysm repair include the need for concurrent aortic valve repair secondary to cusp prolapse into the aneurysm carrying an increased risk of mortality [40, 45, 53].

Yasuda et al. reported freedom from postoperative grade 3 or worse AR at 10 years to be 93% [43]. Surgical procedures included plication (47%), patch repair (40%) or aortic root replacement (13%). They noted a higher incidence of late postoperative complications in patients needing aortic valve replacement. These included: prosthetic valve malfunction, endocarditis, recurrence of aneurysm and thrombosis (0.8%). They recommended early aggressive treatment of endocarditis [40]. In the review of patients by Kuriakose et al., the need for repeat surgical intervention for aneurysm recurrence was reported at 3.2%, with 4.2% requiring re-operation for AR; however, 6.8% had developed new or progressive AR on follow-up [58••].

Percutaneous Closure

Since Cullen et al. attempted the first PC of a SOVA using a Rashkind umbrella in 1994, the less invasive approach has become a viable treatment option for this defect [59]. Since this novel procedure, many other devices have been employed including: ADO [60], Amplatzer septal occlude, PDA and VSD duct occluders (Shanghai memory alloy) [61, 62], Cocoon ductal occluder [63] and Gianturco coils [64] (Table 1). Kuriakose et al. noted that most operators prefer devices with a double disk design. The approach can be

Table 1 Summary of percutaneous device use in previous studies

Study group	Device used	Number	Margins (mm)	Imaging	Defect size (mm)	Percentage success
Guan et al	PDA ductal occluder	10	2–4	TTE	7–15	100
	VSD occluder (shanghai memory alloy)		3–5			
Sinha et al	Cocoon ductal occluder	8	2–4	TEE	9–17	100
Chang et al	Amplatzer ductal occluder	3	2–4	TEE	4–8	100
	Gianturco coil	1				
Kerkar et al	Amplatzer ductal occluder	20	2–4	TEE	4–11	90
Arora et al	Amplatzer ductal occluder	5		TTE	7–12	86
	Amplatzer septal occluder	1				
	Rashkind umbrella	2				
Xiao	VSD occluder	18	2–7	TTE	4–10	96.7
	PDA occluder	11				

retrograde arterial or transvenous and the device should be 2–4 mm larger than the defect [58••].

The procedure is most commonly performed under general anaesthesia. Access is obtained via the femoral vein and artery. The use of heparin (100 units/kg) is recommended during insertion of the device [60, 65••] to reduce the potential for thrombus formation and stroke, and antibiotics (i.e. Cefazolin) should be given to reduce the risk of peri-procedural IE [60]. In order to visualise the defect during intervention, several groups have utilised TTE [65••, 66]. Other groups including Kerkar et al. have preferred TEE guidance, believing that the procedure was made "more comfortable" and allowed them to avoid the need for balloon sizing of the defect [60, 66]. Techniques for device insertion include the use of an Amplatz gooseneck snare to create an exteriorised guidewire loop [61].

Complications from percutaneous device occlusion include the risk of new onset AR, haemolysis from residual shunt and myocardial infarction [61]. One group suggested that selective coronary angiography should be performed to exclude coexisting coronary artery disease or encroachment of the occluder on the coronary ostia. To avoid haemolysis, Arora et al. emphasise the importance of complete occlusion at the time of device insertion [66]. Post-procedural thrombosis prophylaxis with aspirin 3–5 mg/kg for 6 months is recommended following device insertion [61]. Contraindications to PC are large defect size, moderate to severe AR, fistulous aneurysmal tract, presence of IE or congenital anomaly.

Comparison of Surgical Versus Percutaneous Transcatheter Repair

A large systematic review by Kuriakose et al. was recently performed and included 34 studies and 877 patients treated for sinus of Valsalva aneurysm between 1957 and 2014. They compare outcomes in those treated with a catheter guided versus a surgical approach. They noted that comparison between the groups was challenging as patients with more complicated lesions including those with IE, bicuspid aortic valves, tunnel type narrowing, larger defects or multiple ruptures sites were preferentially managed surgically. Although they did note that the New York Heart Association class and Qp:Qs mean 2.3 (range 1.2 to 4.3) was similar between the catheter and surgical closure groups, 60 versus 52% class 3 to 4. They concluded that patients too unwell to tolerated bypass runs, with mild or less AR and more simple associated lesions (ASD, VSD, PDA) can be safely and effective managed with device closure [58••]. Other groups have suggested that PC may be a viable option in high-risk surgical patients [66, 67].

A non-randomised study by Xiao et al. in 2017 enrolled 85 patients with SOVA who underwent repair, including 30 with PC and 55 with surgical repair. Surgical repair was undertaken with a patch technique. Device closure was performed with Shanghai produced VSD and PDA occluders [65••]. A true comparison cannot be made as the groups were selected into each arm based on enrolment criteria. They did, however, state that the PC group showed "no IE, residual shunt, thrombosis, occluder abscission, or other serious complications. Good long-term outcomes and lower complication rates were found compared to the surgical group". However, they also recommend that if there are significant complicating lesions, a surgical approach should be performed [65••]. The advantages of PC lie in minimal invasiveness, a shorter time of hospital stay, an avoidance of extracorporeal circulation and blood transfusion, and potentially economic benefits compared with surgical repair [68]. PC procedural times have been described in eight patients at 42 min with in hospital stay at 2.9 days [67].

Conclusions

Following rupture, sinus of Valsalva aneurysms may result in the rapid development of CCF due to a large volume L–R shunt. TTE is diagnostic in most cases. Surgical repair yields

good long-term results, although in select patient's percutaneous closure may avoid the need for sternotomy and bypass, reducing recovery times. Debate still exists regarding the management of unruptured defects in asymptomatic patients.

Compliance with Ethical Standards

Conflict of Interest Colm R. Breatnach and Kevin P. Walsh declare that they have no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

References

Papers of particular interest, published recently, have been highlighted as:
- • Of importance
- •• Of major importance

1. Perry EL, Burchell HB, Edwards JE. Congenital communication between the left ventricle and the right atrium; co-existing ventricular septal defect and double tricuspid orifice. Proc Staff Meet Mayo Clinic. 1949;24(8):198–206.
2. Gerbode F, Hultgren H, Melrose D, Osborn J. Syndrome of left ventricular-right atrial shunt; successful surgical repair of defect in five cases, with observation of bradycardia on closure. Ann Surg. 1958;148(3):433–46.
3. Sakakibara S, Konno S. Left ventricular-right atrial communication. Ann Surg. 1963;158:93–9.
4. Sinisalo JP, Sreeram N, Jokinen E, Qureshi SA. Acquired left ventricular-right atrium shunts. Eur J Cardiothorac Surg. 2011;39(4):500–6.
5. Yuan SM. Left ventricular to right atrial shunt (Gerbode defect): congenital versus acquired. Postepy Kardiol Interwencyjnej. 2014;10(3):185–94.
6. Lee SY, Song JY, Baek JS. Percutaneous closure of the acquired gerbode shunt using the amplatzer duct occluder in a 3-month old patient. Kor Circ J. 2013;43(6):429–31.
7. Prifti E, Ademaj F, Baboci A, Demiraj A. Acquired Gerbode defect following endocarditis of the tricuspid valve: a case report and literature review. J Cardiothorac Surg. 2015;10:115.
8. Borkar Y, Nayak K, Shetty RK, Bhat G, Moka R. Gerbode ventricular septal defect -a rare cardiac anomaly associated with genetic variants in Indian population- a case series. J Clin Diagn Res. 2017;11(3):GR01–GR4.
9. Gosciniak P, Larysz B, Baraniak J, Jozwa R, Zych A, Mokrzycki K, et al. The Gerbode defect - a frequent echocardiographic pitfall. Kardiol Pol. 2012;70(11):1191–3.
10. •• Yuan SM. A Systematic Review of Acquired Left Ventricle to Right Atrium Shunts (Gerbode Defects). Hellenic J Cardiol. 2015;56(5):357–72. **Yuan reviews 234 patients, comparing outcomes of surgical and percutaneous closure. Insight into patient selection and the advantages of each approach is discussed.**
11. Wasserman SM, Fann JI, Atwood JE, Burdon TA, Fadel BM. Acquired left ventricular-right atrial communication: Gerbode-type defect. Echocardiography. 2002;19(1):67–72.
12. Can I, Krueger K, Chandrashekar Y, Li JM, Dykoski R, Tholakanahalli VN. Images in cardiovascular medicine. Gerbode-type defect induced by catheter ablation of the atrioventricular node. Circulation. 2009;119(22):e553–6.
13. Martinez MW, Mookadam F, Sun Y, Hagler DJ. Transcatheter closure of ischemic and post-traumatic ventricular septal ruptures. Catheter Cardiovasc Interv. 2007;69(3):403–7.
14. Katta S, Akosah K, Stambler B, Salter D, Guerraty A, Mohanty PK. Atrioventricular fistula: an unusual complication of endomyocardial biopsy in a heart transplant recipient. J Am Soc Echocardiogr. 1994;7(4):405–9.
15. Silbiger JJ, Kamran M, Handwerker S, Kumar N, Marcali M. The Gerbode defect: left ventricular to right atrial communication-anatomic, hemodynamic, and echocardiographic features. Echocardiography. 2009;26(8):993–8.
16. Taskesen T, Prouse AF, Goldberg SL, Gill EA. Gerbode defect: another nail for the 3D transesophagel echo hammer? Int J Cardiovasc Imaging. 2015;31(4):753–64.
17. Barclay RS, Reid JM, Coleman EN, Stevenson JG, Welsh TM, McSwan N. Communication between the left ventricle and right atrium. Thorax. 1967;22(5):473–7.
18. Frigg C, Cassina T, Siclari F, Mauri R. Unusual complication after aortic valve replacement. Interact Cardiovasc Thorac Surg. 2008;7(1):149–50.
19. Hansalia S, Manda J, Pothineni KR, Nanda NC. Usefulness of live/real time three-dimensional transthoracic echocardiography in diagnosing acquired left ventricular-right atrial communication misdiagnosed as severe pulmonary hypertension by two-dimensional transthoracic echocardiography. Echocardiography. 2009;26(2):224–7.
20. Mahajan K, Negi P, Asotra S, Rana V. Infravalvular type of Gerbode defect: a rare cardiac anomaly. BMJ Case Rep. 201; https://doi.org/10.1136/bcr-2015-214091.
21. Trehan V, Ramakrishnan S, Goyal NK. Successful device closure of an acquired Gerbode defect. Catheter Cardiovasc Interv. 2006;68(6):942–5.
22. Mousavi N, Shook DC, Kilcullen N, Aranki S, Kwong RY, Landzberg MJ, et al. Multimodality imaging of a Gerbode defect. Circulation. 2012;126(1):e1–2.
23. Yacoub MH, Mansur A, Towers M, Westbury H. Bacterial endocarditis complicating left ventricle to right atrium communication. Br J Dis Chest. 1972;66(1):78–82.
24. Toprak C, Kahveci G, Akpinar S, Tabakci MM, Guler Y. Concomitant Gerbode-like defect and anterior mitral leaflet perforation after aortic valve replacement for endocarditis. Echocardiography. 2013;30(8):E231–5.
25. Hsu SY, Shen TC. A spontaneously closed, acquired supravalvular Gerbode defect mimicking an unruptured sinus of Valsalva aneurysm. Eur Heart J Cardiovasc Imaging. 2014;15(4):471.
26. Wu MH, Wang JK, Lin MT, Wu ET, Lu FL, Chiu SN, et al. Ventricular septal defect with secondary left ventricular-to-right atrial shunt is associated with a higher risk for infective endocarditis and a lower late chance of closure. Pediatrics. 2006;117(2):e262–7.
27. Ho JO, Moghrabi F, Deshpande RP. Group B Streptococcus endocarditis with left ventricle-right atrium (Gerbode's) defect. Ann Thorac Surg. 2014;97(6):e165–6.
28. Velebit V, Schoneberger A, Ciaroni S, Bloch A, Maurice J, Christenson JT, et al. "Acquired" left ventricular-to-right atrial shunt (Gerbode defect) after bacterial endocarditis. Tex Heart Inst J. 1995;22(1):100–2.
29. Lorber A, Nair P, Gruberg L. Transcatheter closure of acquired Gerbode defect following mitral valve replacement using the Amplatzer duct occluder. J Invasive Cardiol. 2006;18(10):E264–6.
30. Rothman A, Galindo A, Channick R, Blanchard D. Amplatzer device closure of a tortuous Gerbode (left ventricle-to-right atrium) defect complicated by transient hemolysis in an octogenarian. J Invasive Cardiol. 2008;20(9):E273–6.

31. Newman JN Jr, Rozanski L, Kreulen T. Acquired left ventricular to right atrial intracardiac shunt after myocardial infarction: a case report and review of the literature. J Am Soc Echocardiogr. 1996;9(5):716–20.
32. Ganesan G, Paul GJ, Mahadevan VS. Transcatheter closure of left ventricle to right atrial communication using cera duct occluder. Indian Heart J. 2017;69(3):341–4.
33. Phan QT, Kim SW, Nguyen HL. Percutaneous closure of congenital Gerbode defect using Nit-Occlud((R)) Le VSD coil. World J Cardiol. 2017;9(7):634–9.
34. Sinisalo J, Sreeram N, Qureshi SA. Transcatheter closure of acquired left ventricle to right atrium shunts. Catheter Cardiovasc Interv. 2013;82(6):E809–14.
35.• Vijayalakshmi IB, Natraj Setty HS, Chitra N, Manjunath CN. Amplatzer duct occluder II for closure of congenital Gerbode defects. Catheter Cardiovasc Interv. 2015;86(6):1057–62. **A recent report on the ADO II device in 12 patients aged 10 months to 16 years, describing excellent results.**
36. Thurnam J. On aneurisms, and especially spontaneous varicose aneurisms of the ascending aorta, and sinuses of Valsalva, with cases. Med Chir Trans. 1840;23:323–84.
37. Edwards JE, Burchell HB. The pathological anatomy of deficiencies between the aortic root and the heart, including aortic sinus aneurysms. Thorax. 1957;12(2):125–39.
38. Lillehei CW, Stanley P, Varco RL. Surgical treatment of ruptured aneurysms of the sinus of Valsalva. Ann Surg. 1957;146(3):459–72.
39. Sivadasanpillai H, Valaparambil A, Sivasubramonian S, Mahadevan KK, Sasidharan B, Namboodiri N, et al. Percutaneous closure of ruptured sinus of Valsalva aneurysms: intermediate term follow-up results. EuroIntervention. 2010;6(2):214–9.
40. Takach TJ, Reul GJ, Duncan JM, Cooley DA, Livesay JJ, Ott DA, et al. Sinus of Valsalva aneurysm or fistula: management and outcome. Ann Thorac Surg. 1999;68(5):1573–7.
41. Chu SH, Hung CR, How SS, Chang H, Wang SS, Tsai CH, et al. Ruptured aneurysms of the sinus of Valsalva in oriental patients. J Thorac Cardiovasc Surg. 1990;99(2):288–98.
42. Dong C, Wu QY, Tang Y. Ruptured sinus of valsalva aneurysm: a Beijing experience. Ann Thorac Surg. 2002;74(5):1621–4.
43. Murashita T, Kubota T, Kamikubo Y, Shiiya N, Yasuda K. Long-term results of aortic valve regurgitation after repair of ruptured sinus of valsalva aneurysm. Ann Thorac Surg. 2002;73(5):1466–71.
44. Zikri MA, Stewart RW, Cosgrove DM. Surgical correction for sinus of Valsalva aneurysm. J Cardiovasc Surg. 1999;40(6):787–91.
45. Moustafa S, Mookadam F, Cooper L, Adam G, Zehr K, Stulak J, et al. Sinus of Valsalva aneurysms–47 years of a single center experience and systematic overview of published reports. Am J Cardiol. 2007;99(8):1159–64.
46. Goldberg N, Krasnow N. Sinus of Valsalva aneurysms. Clin Cardiol. 1990;13(12):831–6.
47. Azakie A, David TE, Peniston CM, Rao V, Williams WG. Ruptured sinus of valsalva aneurysm: early recurrence and fate of the aortic valve. Ann Thorac Surg. 2000;70(5):1466–70. **discussion 70-1**
48. Rao PS, Bromberg BI, Jureidini SB, Fiore AC. Transcatheter occlusion of ruptured sinus of valsalva aneurysm: innovative use of available technology. Catheter Cardiovasc Interv. 2003;58(1):130–4.
49. Shahrabani RM, Jairaj PS. Unruptured aneurysm of the sinus of Valsalva: a potential source of cerebrovascular embolism. Br Heart J. 1993;69(3):266–7.
50. Herrador JA, Aragon V, Fernandez JC. Thrombosis of a Valsalva sinus aneurysm causing AMI. Rev Esp Cardiol. 2018;71(1):44.
51. Umeda H, Isotani A, Arita T, Kudo T, Matsunaga T, Otsuji Y, et al. Rapid growth of thrombus formation in the unruptured sinus of Valsalva aneurysm following coronary angiography. J Echocardiogr. 2018; https://doi.org/10.1007/s12574-018-0378-4.
52. Abe T, Komatsu S. Surgical repair and long-term results in ruptured sinus of Valsalva aneurysm. Ann Thorac Surg. 1988;46(5):520–5.
53. Wang ZJ, Zou CW, Li DC, Li HX, Wang AB, Yuan GD, et al. Surgical repair of sinus of Valsalva aneurysm in Asian patients. Ann Thorac Surg. 2007;84(1):156–60.
54. Vural KM, Sener E, Tasdemir O, Bayazit K. Approach to sinus of Valsalva aneurysms: a review of 53 cases. Eur J Cardiothoracic Surg. 2001;20(1):71–6.
55. van Son JA, Danielson GK, Schaff HV, Orszulak TA, Edwards WD, Seward JB. Long-term outcome of surgical repair of ruptured sinus of Valsalva aneurysm. Circulation. 1994;90(5 Pt 2):II20–9.
56. Taguchi K, Sasaki N, Matsuura Y, Uemura R. Surgical correction of aneurysm of the sinus of Valsalva. A report of forty-five consecutive patients including eight with total replacement of the aortic valve. Am J Cardiol. 1969;23(2):180–91.
57. Au WK, Chiu SW, Mok CK, Lee WT, Cheung D, He GW. Repair of ruptured sinus of valsalva aneurysm: determinants of long-term survival. Ann Thorac Surg. 1998;66(5):1604–10.
58.•• Kuriakose EM, Bhatla P, McElhinney DB. Comparison of reported outcomes with percutaneous versus surgical closure of ruptured sinus of Valsalva aneurysm. Am J Cardiol. 2015;115(3):392–8. **This review assesses a large historical cohort of patients (877) with SOVA over a 57 year period and provides insight into recent advances in imaging and management.**
59. Cullen S, Somerville J, Redington A. Transcatheter closure of a ruptured aneurysm of the sinus of Valsalva. Br Heart J. 1994;71(5):479–80.
60. Kerkar PG, Lanjewar CP, Mishra N, Nyayadhish P, Mammen I. Transcatheter closure of ruptured sinus of Valsalva aneurysm using the Amplatzer duct occluder: immediate results and mid-term follow-up. Eur Heart J. 2010;31(23):2881–7.
61. Guan L, Zhou D, Zhang F, Pan W, Dong L, Chen H, et al. Percutaneous device closure of ruptured sinus of valsalva aneurysm: a preliminary experience. J Invasive Cardiol. 2013;25(10):492–6.
62. Cronin EM, Walsh KP, Blake GJ. Ruptured noncoronary sinus of valsalva aneurysm: an unusual cause of chronic right-sided heart failure. Circ Heart Fail. 2011;4(1):e3–4.
63. Sinha SK, Khanna NN, Razi M, Krishna V, Jha MJ, Mishra V, et al. Safety and feasibility of Transcatheter interruption of ruptured sinus of Valsalva aneurysm using the cocoon duct Occluder: immediate results and mid-term follow-up. Cardiol Res. 2017;8(4):154–60.
64. Chang CW, Chiu SN, Wu ET, Tsai SK, Wu MH, Wang JK. Transcatheter closure of a ruptured sinus of valsalva aneurysm. Circ J. 2006;70(8):1043–7.
65.•• Xiao JW, Wang QG, Zhang DZ, Cui CS, Han X, Zhang P, et al. Clinical outcomes of percutaneous or surgical closure of ruptured sinus of Valsalva aneurysm. Congenit Heart Dis. 2018;13(2):305–10. **This a large study comparing surgical and percutaneous closure of SOVA in 88 patient, highlighting the advantages and pitfalls of each approach.**
66. Arora R, Trehan V, Rangasetty UM, Mukhopadhyay S, Thakur AK, Kalra GS. Transcatheter closure of ruptured sinus of valsalva aneurysm. J Interv Cardiol. 2004;17(1):53–8.
67. Sen S, Chattopadhyay A, Ray M, Bandyopadhyay B. Transcatheter device closure of ruptured sinus of Valsalva: immediate results and short term follow up. Ann Pediatr Cardiol. 2009;2(1):79–82.
68. Liu S, Xu X, Ding X, Liu G, Zhao Z, Zhao X, et al. Comparison of immediate results and mid-term follow-up of surgical and percutaneous closure of ruptured sinus of Valsalva aneurysm. J Cardiol. 2014;63(3):239–43.