

# Acquired aortocameral fistula occurring late after infective endocarditis: An emblematic case and review of 38 reported cases

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## Abstract

### AIM

To delineate the features and current therapeutic option of congenital and acquired aortocameral fistulas (ACF) secondary to iatrogenic or infectious disorders.

### METHODS

From a PubMed search using the term "aortocameral fistula", 30 suitable papers for the current review were retrieved. Reviews, case series and case reports published in English were considered. Abstracts and reports from scientific meetings were not included. A total of 38 reviewed subjects were collected and analyzed. In addition, another case - an adult male who presented with ACF between commissures of the right and non-coronary sinuses and right atrium as a late complication of *Staphylococcus aureus* infective endocarditis of the AV - is added, the world literature is briefly reviewed.

### RESULTS

A total of thirty-eight subjects producing 39 fistulas were reviewed, analyzed and stratified into either congenital (47%) or acquired (53%) according to their etiology. Of all subjects, 11% were asymptomatic and 89% were symptomatic with dyspnea (21 ×) as the most common presentation. Diagnosis was established by a multidagnostic approach in 23 (60%), single method in 14 (37%) (echocardiography in 12 and catheterization in 2), and at autopsy in 2 (3%) of the subjects. Treatment options included percutaneous transcatheter closure in 12 (30%) with the deployment of the Amplatzer duct or septal occluder and Gianturco coil and surgical correction in 24 (63%).

### CONCLUSION

Acquired ACF is an infrequent entity which may occur late after an episode of endocarditis of the native AV. The management of ACF is generally by surgical correction but non-surgical device intervention has recently been introduced as a safe alternative.

**Key words:** Aortic-atrial shunt; Aortic-atrial fistulas; Infective endocarditis; Late complication; Surgical correction

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**Core tip:** Aortocameral fistula is an uncommon complication of native aortic valve (AV) endocarditis, which is associated with high morbidity and mortality. Acquired aortocameral fistulas (ACF) may originate from any of the three sinuses of Valsalva. Audible continuous murmur may raise suspicion for the presence of ACF. Congenital fistulas are less commonly reported than the acquired types. Acquired ACF may occur late after an episode of endocarditis of the native AV. The management of ACF is generally by surgical correction but non-surgical device intervention has recently been introduced as a safe alternative. Another case is added and the world literature is briefly reviewed.

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## INTRODUCTION

Aortocameral fistulas (ACF) may be congenital<sup>[1]</sup> or acquired complicating acute aortic dissection<sup>[2]</sup> following an intimal tear in the vicinity and proximity of the aortic root or after aortic valve (AV) replacement<sup>[3]</sup>. ACF is an uncommon complication of native AV endocarditis, which is associated with high morbidity and mortality. ACF may originate from any of the three sinuses of Valsalva. Audible continuous murmur may raise suspicion for the presence of ACF<sup>[4]</sup>. The clinical manifestations of ACF may include exertional dyspnea<sup>[2,5]</sup>, chest pain<sup>[6,7]</sup>, palpitation<sup>[6,8]</sup>, congestive heart failure<sup>[9,10]</sup> and recurrent respiratory tract infection<sup>[11,12]</sup>. ACF may incidentally be found during routine preoperative examination<sup>[13]</sup>. Untreated ACF may cause significant morbidity and early mortality. The surgical correction of ACF is the treatment of choice but percutaneous transcatheter device intervention has recently been successfully introduced for the closure of ACF<sup>[5,6,8,9]</sup>. Acquired ACF is an infrequent entity which may occur late after an episode of endocarditis of the native AV. Another case of our own is added and the world literature is briefly reviewed.

## MATERIALS AND METHODS

### Literature search

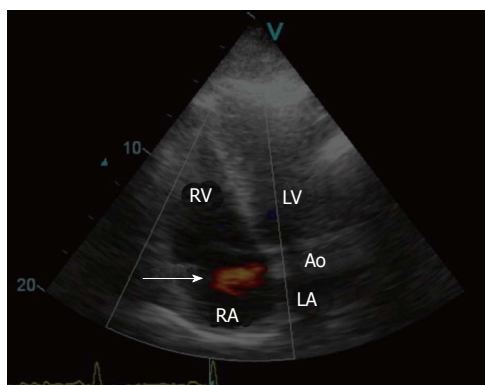
From the PubMed search using the term "aortocameral fistula", 30 suitable papers for the current review were retrieved (Table 1). Reviews, case series and

case reports published in English were considered. Abstracts and reports from scientific meetings were not included. From 30 publications, 38 reviewed subjects were collected and analyzed. Data were analyzed using descriptive statistics.

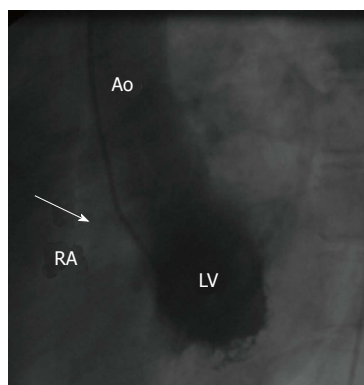
### Additional clinical case

An adult male presented with ACF between the junction of RCS-NCS and RA as a late complication of *Staphylococcus aureus* infective endocarditis (IE) of the native AV, is added.

A 44-year-old male survivor of a prior episode of *Staphylococcus aureus* IE of the native AV (1998) presented with a recent history of rapid fatigability (2008) during sporting activities. He was afebrile and a continuous murmur was heard. Laboratory results and chest X-ray were normal. Resting ECG depicted sinus rhythm with signs of left ventricular hypertrophy (LVH). Two-dimensional transthoracic Doppler echocardiography revealed mild LVH, the right ventricle (RV) was dilated and normokinetic, and the tricuspid AV had no vegetation. Color flow mapping revealed evidence of a high velocity shunt between the commissures of the right coronary sinus (RCS) and non-coronary sinuses (NCS) terminating into the right atrium (RA) (Figure 1, Supplementary material online, Video 1). Cardiac catheterization demonstrated a shunt between the aorta and the RA and normal left ventricular kinetics (Figure 2, Supplementary material online, Video 2). Hemodynamic evaluation revealed a significant left-to-right shunt (Qp: Qs = 2.0:1.0) with normal pulmonary vascular resistance, normal intracardiac pressures and high resting cardiac output of 10 L/min. Computed tomography and cardiovascular magnetic resonance were not available at that time. The fistula was surgically closed (2008). The fistula was surgically closed (2008). After establishing median sternotomy, extracorporeal circulation was performed through standard cannulation of the aorta and right atrium. The heart was arrested with antegrade and selective blood cardioplegia. On inspection, no infectious masses or evidence of abscess or vegetations were visible. Further inspection revealed that the ascending aorta was not dilated or calcified and the LV showed moderate hypertrophy. After aortotomy, the AV could be inspected, which was tricuspid with mild thickening and the fistula was clearly visible between the RCS and NCS terminating into RA. The fistula was closed with 4.0 prolene suture and pledgets. The patient could easily be weaned off after an uneventful procedure. Postoperative transesophageal echocardiography revealed no rest shunt flow. The patient had an uneventful postoperative course. The patient had uneventful postoperative course and regained his non-professional sporting activities without any limitations. After 8 years of follow-up, he remains free of symptoms. The fistula was closed by 4.0 prolene suture and pledgets. The patient had uneventful postoperative course and regained his non-professional sporting activities without any limitations. After 7 years of



**Figure 1** A frame of colored Doppler trans-thoracic echocardiography, five-chamber view illustrating the aortic-atrial fistula (arrow). Ao: Aorta; RA: Right atrium; LA: Left atrium; RV: Right ventricle; LV: Left ventricle.



**Figure 2** Levo-ventriculogram in the left anterior oblique view demonstrating the fistula (arrow) between the right and non-coronary sinuses communicating with the right atrium. RCS/NCS: Right and non-coronary sinuses; LV: Levo-ventriculogram; RA: Right atrium.

follow-up, he remains free of symptoms.

### Statistical analysis

In contrast to classic meta-analysis, the outcome is defined here as the percentages of an event (without comparison) in observed patients.

## RESULTS

A total of 38 subjects were reviewed [21 males (55%) and 17 females (45%)], with a mean age of 36.8 years (range 3-70 years). The etiology was congenital in 18 (47%) and acquired in 20 (53%). They all had 39 fistulas. Of those, 37 had a single and 1 had dual origin, with a similar outflow distribution of 37 single and one dual termination. Their origin was NCS in 21 (54%), RCS in 10 (26%), left coronary sinus (LCS) in 4 (10%) and the thoracic aorta in 4 (10%) of the subjects. The termination was into RA in 29 (74%), left atrium (LA) in 6 (15%), RV in 2 (5%), pulmonary artery in 1 (3%) and right ventricular outflow tract in 1 (3%). Four subjects (11%) were asymptomatic. In the symptomatic subjects (89%), the most common presentations were dyspnea (21 ×), followed by congestive heart failure (6 ×), chest pain (7 ×), palpitations (7 ×), IE (6 ×) with *Streptococcus mitis*<sup>[14]</sup> and *Staphylococcus epidermidis*<sup>[15]</sup>. Although syncope, hematuria, hemoptysis and recurrent respiratory tract infection are rarely reported, sudden death has also been observed<sup>[16]</sup>.

Diagnosis was established by multidagnostic approach in 23 (60%), a single method in 14 (37%) (echocardiography in 12 and catheterization in 2), and at autopsy in 2 (3%)<sup>[17]</sup> of the subjects. IE was found in 6 (16%) subjects, all originating from the NCS and communicating with the RA in 5 and the LA in one. Treatment included percutaneous transcatheter closure in 12 (30%) and surgical correction in 24 (63%) and there were 3 mortalities (7%) (Table 1).

## DISCUSSION

An aortic-atrial fistula is an aortocameral fistula pre-

senting as an extracardiac vascular communication that may be congenital<sup>[1]</sup> or acquired<sup>[2]</sup>. ACF is an uncommon complication of native AV endocarditis, which is associated with high morbidity and mortality. In 1963, Kuipers *et al.*<sup>[18]</sup> reported spontaneous ruptured aortic dissection into the right atrium. Congenital fistulas are less commonly reported than the acquired types. ACF may originate from any of the 3 sinuses of Valsalva. Acquired ACF may occur following bacterial endocarditis<sup>[19]</sup>, acute aortic dissection<sup>[2]</sup>, ruptured sinus of Valsalva aneurysm (RSVA)<sup>[19]</sup>, and post-cardiovascular surgical procedures associated with<sup>[5]</sup> or without infective endocarditis (IE)<sup>[20-22]</sup>. Furthermore, ACF may occur after coronary artery bypass grafting<sup>[23]</sup>, after mitral valve replacement<sup>[23]</sup>, following repeat AV replacement<sup>[24]</sup> or secondary to iatrogenic endovascular injury during an invasive diagnostic procedure<sup>[4]</sup>.

ACF may incidentally be found during routine pre-operative examination<sup>[13]</sup> or presented with severe heart failure<sup>[2]</sup>. The surgical correction of ACF is the treatment of choice but percutaneous transcatheter device intervention has recently been successfully introduced for the closure of ACF<sup>[5,6,8,9]</sup>.

In 1831, Hope<sup>[25]</sup> described a ruptured aneurysm of a sinus of Valsalva into the right atrium. Congenital or acquired aortic-atrial fistulas are rare anomalies. In 1924, a large autopsy series ( $n = 4000$ ) revealed aorta to atrium fistula as an incidental finding; rupture was found in 1197, of which 13 were into the RA due to infectious, traumatic and atherosclerotic causes<sup>[26]</sup>. ACF may be congenital<sup>[1]</sup> or acquired complicating acute aortic dissection<sup>[2]</sup> following an intimal tear in the vicinity and proximity of the aortic root or after AV replacement<sup>[3]</sup> (Table 2). ACF may occur in patients with infective endocarditis<sup>[19]</sup>, as was the case in the current patient and in 16% of the reviewed subjects.

### Clinical presentation

Audible continuous murmur may raise suspicion for the presence of ACF<sup>[4]</sup>. The clinical manifestations of ACF may include exertional dyspnea<sup>[2,5]</sup>, chest pain<sup>[6,7]</sup>, palpitation<sup>[6,8]</sup>, congestive heart failure<sup>[9,10]</sup> and recurrent

Table 1 Clinical presentations and management of 39 subjects

Ref.	Age gender	ACF	Diagnostic modality	Clinical presentation/etiology	Management
Jung <i>et al</i> <sup>[19]</sup> , 2011	49 M	NCS-right atrium	TTE	Dyspnea and high fever Ruptured sinus of Valsalva Infective endocarditis ( <i>Enterococcus gallinarum</i> )	Patch repair AVR/TVR
Raufi <i>et al</i> <sup>[21]</sup> , 2002	50 M	RCS-right atrium	TEE aortography cardiac cath	Dyspnea and chest pain Post-repair of aneurysm of right sinus of Valsalva	Repair of the ruptured sinus of Valsalva Closure of the fistula
Hsu <i>et al</i> <sup>[2]</sup> , 2000	67 M	False lumen-Right atrium	TTE cardiac cath ioTEE	Dyspnea Acute aortic dissection	Fistula repair Bentall procedure
Chung <i>et al</i> <sup>[20]</sup> , 2000	52 M	AAR-right atrium	TTE cardiac cath angiography MRI	Dyspnea and hemoptysis Post-repair (ARR) of acute aortic dissection	Closure of the fistula New composite aortic root graft
Ananthasubramaniam <i>et al</i> <sup>[24]</sup> , 2005	66 M	LCS-left atrium	TTE TEE ioTEE	Dyspnea post-AVR	Surgical closure of the fistula/repair
Haddad <i>et al</i> <sup>[22]</sup> , 2008	66 M	NCS-right atrium	TTE	Dyspnea post-repair of acute aortic dissection	Fistula repair Bentall procedure
Estévez-Loureiro <i>et al</i> <sup>[5]</sup> , 2012	44 M	NCS-left atrium	TTE TEE MDCT CAG 3-D TEE	Dyspnea Infective endocarditis post-AVR ( <i>Streptococcus viridans</i> )	Percutaneous Amplatzer vascular plug III occluder
Bouchez <i>et al</i> <sup>[13]</sup> , 2012	61 M	LCS-left atrium	io TEE 3-D TEE	Asymptomatic	Conservative
Mundo-Sagardía <i>et al</i> <sup>[14]</sup> , 2006	22 M	NCS-right atrium	TTE TEE	Infective endocarditis ( <i>Streptococcus mitis</i> ). Complex congenital heart disease. Perforation of sinus Valsalva aneurysm (NCS)	Closure/repair
Ladowski <i>et al</i> <sup>[4]</sup> , 1984	56 F	NCS-right atrium	angiography	Iatrogenic dissection	Surgical closure/repair
Vydt <i>et al</i> <sup>[16]</sup> , 2002	43 M	NCS-right atrium	TTE TEE cardiac cath angiography aortography	Chest pain, DOE, ruptured sinus Valsalva aneurysm	Surgical closure
Moiduddin <i>et al</i> <sup>[12]</sup> , 2009	5 F	NCS-left atrium	TEE angiography cardiac cath	Amplatzer atrial septal occluder ASD PDA	Surgical closure/repair
Chandra <i>et al</i> <sup>[11]</sup> , 2011	12 F	RCS-right atrium	TTE CTA aortography angiography cardiac cath	Dyspnea, palpitation	Percutaneous Amplatzer duct occluder
Noureddine <i>et al</i> <sup>[16]</sup> , 2001	21 F	NCS-right atrium	TTE TEE	Dyspnea	Sudden death
Przybojewski <i>et al</i> <sup>[39]</sup> , 1983	27 M	RCS/right atrium and right ventricle	Phonocardiography TTE cardiac cath angiography aortography	Dyspnea; biventricular heart failure, ruptured sinus Valsalva aneurysm (RCS)	Surgical closure/repair
Mujanovic <i>et al</i> <sup>[35]</sup> , 2010	41 F	NCS-right atrium	TTE angiography	Heart failure; ruptured sinus Valsalva aneurysm (NCS)	Surgical closure/repair
Mello <i>et al</i> <sup>[33]</sup> , 2005	16 F	NCS-left atrium	TTE TEE aortography	Asymptomatic, post-placement of Amplatzer atrial septal occluder (ASO) for ASDII	Surgical closure/repair
Grayburn <i>et al</i> <sup>[7]</sup> , 2005	41 F	Aorta-right atrium	TTE TEE	Chest pain, post-placement of Amplatzer atrial septal occluder (ASO) for ASD	Surgical closure/repair
Chun <i>et al</i> <sup>[30]</sup> , 2003	10 M	NCS-right atrium	TTE TEE	Asymptomatic, post-placement of Amplatzer atrial septal occluder (ASO) for ASD	Surgical closure/repair
Knirsch <i>et al</i> <sup>[32]</sup> , 2005	3 M	NCS-left atrium	TTE	Asymptomatic, post-placement of Amplatzer atrial septal occluder (ASO) for ASD	Surgical closure/repair
Jang <i>et al</i> <sup>[31]</sup> , 2005	54 F	NCS-right atrium	TTE	Dyspnea, palpitation, hematuria, post-placement of Amplatzer atrial septal occluder (ASO) for ASDII	Surgical closure/repair
Ozay <i>et al</i> <sup>[45]</sup> , 2007	22 F	NCS-right atrium	TTE TEE aortography cardiac catheter	Palpitation post-surgical repair of VSD and ASDII	Surgical closure/repair/correction
Elwatidy <i>et al</i> <sup>[46]</sup> , 2003	3 F	Aortic isthmus-RA	TTE cardiac catheter	Presented as a case of PDA	Surgical closure/repair/correction
Akowuah <i>et al</i> <sup>[10]</sup> , 2002	52 F	NCS-right atrium	TTE cardiac catheter	CHF, IE, TV, <i>Staphylococcus aureus</i> MRSA	Surgical TVR correction
Darwazah <i>et al</i> <sup>[15]</sup> , 2006	23 M	NCS/LCS-right atrium	TTE	PVE of AVR <i>Staphylococcus epidermidis</i>	Surgical Re-re AVR correction
Russo <i>et al</i> <sup>[47]</sup> , 2001	70 F	NCS-right atrium	TTE TEE	Chest pain, dyspnea, CHF complication of AAD type 1	Surgical closure/repair/correction
Onorato <i>et al</i> <sup>[9]</sup> , 2005	48 F	NCS-right atrium	TTE TEE ICE aortography cardiac catheter	Dyspnea, CHF, ruptured sinus Valsalva aneurysm	ADO catheter closure



Chang <i>et al</i> <sup>[8]</sup> , 2006	47, 22 F (2 ×) and 22, 18 M (2 ×)	NCS-RA (1 ×) and RCS-RA (1 ×) and RCS-RV (2 ×)	4 × TEE 4 × aortography	Closure of VSD and AVR, IE (1 ×) Dyspnea and palpitation (3 ×)	ADO catheter closure (3 ×) and Gianturco coil (1 ×)
Szkutnik <i>et al</i> <sup>[6]</sup> , 2009 congenital (4 ×) and acquired (1 ×)	51, 23, 41 M (3 ×) and 18, 28 F (2 ×)	RCS-RVOT (3 ×), RCS-RA (1 ×) LCS-PA (1 ×) and NCS-RA (1 ×)	TTE TEE MDCT	Dyspnea, chest pain, palpitation and syncope	ADO (5 ×) and ASO (1 ×) catheter closure
Oram <i>et al</i> <sup>[17]</sup> , 1955	36, 67 M (2 ×)	NCS-RA RCS-RA, RV	Catheterization, autopsy	Chest pain, palpitation, dyspnea	Chest pain, Post-mortem
Said and Mariani 2016	44 M	NCS-RCS-RA	TTE, aortography cardiac cath, angiography	Easy fatigability	Surgical closure

AAD: Acute aortic dissection; ACF: Aortocameral fistula; AAR: Aneurysm of the aortic root; ADO: Amplatzer duct occlude; ARR: Aortic root replacement; ASD: Atrial septal defect; ASO: Amplatzer atrial septal occlude; AVR: Aortic valve replacement; CHF: Congestive heart failure; CP: Chest pain; CTA: Computed tomography angiography; D: Dyspnea; DOE: Dyspnea on exertion; F: Female; ICE: Intra-cardiac echocardiography; IE: Infective endocarditis; io: Intra-operative; LCS: Left coronary sinus; M: Male; MDCT: Multi-detector computed tomography; MRI: Magnetic resonance imaging; NCS: Non-coronary sinus; PA: Pulmonary artery; PDA: Patent ductus arteriosus; PVE: Prosthetic valve endocarditis; RA: Right atrium; RCS: Right coronary sinus; RV: Right ventricle; RVOT: Right ventricular outflow tract; TEE: Transesophageal echocardiography; TTE: Transthoracic echocardiography; TV: Tricuspid valve; TVR: Tricuspid valve replacement; VSD: Ventricular septal defect.

Table 2 Etiology of aortocameral fistulas

Etiology	Condition/references
Congenital	Congenital RCS-RA fistula <sup>[1]</sup> and aortic isthmus-RA fistula <sup>[46]</sup>
Acquired-iatrogenic (post-surgical and non-surgical intervention/infectious/diagnostic procedures)	Iatrogenic aorta-right atrial fistula: late (14 years) post-surgical repair of VSD and ASD <sup>[45]</sup>  Post-corrective surgery of sinus of Valsalva aneurysm <sup>[21]</sup> Post-CABG <sup>[23,48]</sup> Post-AVR <sup>[3,8]</sup> Post-MVR <sup>[23]</sup>  Post-ARR, after operating on a type A dissection <sup>[20,47]</sup> Following ASO closure of the secundum ASD II <sup>[30]</sup> NVE <sup>[10]</sup> , RCS/NCS-right atrial fistula (current case) secondary to NVE PVE <sup>[5,15]</sup>  ACF associated with diagnostic cardiac catheterization (NCS-RAA) <sup>[4]</sup> ACF post-non-penetrating thoracic injury <sup>[49]</sup> has been reported RSVA <sup>[27]</sup>  Rupture of ascending aorta aneurysm <sup>[18]</sup>
Acquired-accidental/traumatic	
Spontaneous	

ACF: Aortocameral fistula; ARR: Aortic root replacement; ASD: Atrial septal defect; ASO: Amplatzer atrial septal occlude; AVR: Aortic valve replacement; CABG: Coronary artery bypass grafting; MVR: Mitral valve replacement; NCS: Non-coronary sinus; NVE: Native valve endocarditis; PVE: Prosthetic valve endocarditis; RA: Right atrium; RAA: Right atrial appendage; RCS: Right coronary sinus; RSVA: Ruptured sinus Valsalva aneurysm; VSD: Ventricular septal defect.

respiratory tract infection<sup>[11,12]</sup>. Our patient presented with reduced physical fitness as the only symptom, occurring late after the index native valve endocarditis. Among the 38 reviewed subjects, four (11%) were asymptomatic and the majority (89%) were symptomatic.

ACF may originate from any of the three sinuses of Valsalva, but origin from the NCS was infrequently reported<sup>[27]</sup>. Congenital aneurysms (origin RCS 65%-85%, NCS 10%-30% and LCS < 5%) of the sinus of Valsalva have a tendency to rupture, mainly into the right cardiac chambers (termination RV 63%, RA 32%), resulting in an ACF<sup>[28,29]</sup>. Congenital aneurysms of the sinus of Valsalva may be associated with other defects including bicuspid AV, ventricular septal defect and coarctation of the aorta<sup>[21]</sup>.

ACF may occur between the aorta and right atrium<sup>[2]</sup>, as was the case in our current patient, or left atrium<sup>[24]</sup>.

Congenital ACF may be incidentally found in asymptomatic adult subjects<sup>[13]</sup>. There have been a few reports of iatrogenic acquired fistula formation associated with the percutaneous device closure of atrial septal defects with an Amplatzer septal occluder<sup>[30-33]</sup>.

Congenital aortic-atrial fistulas are extremely rare. Acquired ACF are related to prosthetic valve disorders after aortic root repair associated with<sup>[5,15]</sup> or without infective endocarditis<sup>[22]</sup>. The current patient had a prior IE of the native AV. ACF may appear as an early<sup>[34]</sup>, immediate<sup>[22]</sup> (10 d) or late (4 years)<sup>[5]</sup> postoperative complication.

### Diagnostic modalities

Echocardiography [transthoracic (TTE), transesophageal (TEE) and 3-D TEE]<sup>[7,32,35]</sup> is the first diagnostic modality of choice to precisely delineate the fistula components. With complete right and left cardiac catheterization

and aortography of the aortic root, the fistula can be appropriately evaluated and the exact location indicated<sup>[21,36]</sup>. TTE, TEE and 3-D TEE comprise a useful non-invasive diagnostic modality with which to delineate the fistula characteristics. With 2-D echocardiography, TEE, the clinical diagnosis of ACF may be established but ascending aortography is essential for confirmation and to differentiate from other disorders such as ruptured sinus of Valsalva aneurysm<sup>[36]</sup>, aorta-right atrial tunnel<sup>[11]</sup> and acquired<sup>[37]</sup> or congenital<sup>[38]</sup> coronary cameral fistulas.

A multimodality imaging strategy confirms the diagnosis of ACF. Echocardiography (TTE and TEE), selective coronary angiography and retrograde aortography are used for visualization of the coronary ostia and demonstration of the course of the fistula<sup>[12,36]</sup>. This was the chosen approach in two-third (60%) of the reviewed subjects and in the presented case.

Computed tomography (CT) scan and cardiovascular magnetic resonance imaging (MRI): These diagnostic modalities were not widely applied among the reviewed subjects. In only few cases, CT scan<sup>[1,5,6]</sup> was performed and MRI technique was found in the case reported by Chung<sup>[20]</sup>. In our current case, CT and cardiovascular magnetic resonance were not available at that time and moreover, echocardiography and aortography provided adequate imaging quality of the ACF making further investigations unnecessary.

The most common termination sites of "spontaneously" ruptured aneurysms of coronary sinus of Valsalva are into the RA or RV<sup>[27,39]</sup>; more rarely, the left ventricle<sup>[27]</sup> may be involved, ensuing acute volume overload of the involved cardiac chamber. Our patient had an acquired aortic-right atrial connection.

The origin of congenital aneurysm is generally related to the right coronary sinus (65%-85%)<sup>[1,28,29,39]</sup> and those associated with infective endocarditis ensue from the left coronary sinus<sup>[40]</sup>, RCS<sup>[41]</sup> or NCS<sup>[42]</sup>.

### Management

The first successful surgical correction of ruptured sinus Valsalva aneurysm (RSVA) was reported in 1957<sup>[43]</sup>. In 1966, Temple *et al*<sup>[44]</sup> described the successful surgical repair of aortic-right atrial fistula in an adult symptomatic male. ACF may be closed by surgical intervention<sup>[2]</sup> or by transcatheter device<sup>[5]</sup>. The treatment of choice is early surgical repair, which is necessary to prevent the development of severe symptoms and complications. Untreated ACF may cause significant morbidity and early mortality. Recently, percutaneous transcatheter treatment of ACF has been reported which is considered a novel method for selected cases<sup>[5]</sup>. Percutaneous transcatheter closure of ACF, using the Amplatzer duct occluder, Gianturco coil or Amplatzer septal occluder, has proven to be a safe technique which is gaining territory in the non-surgical management of ACF<sup>[5,6,8,9]</sup>. Our patient had a successful surgical repair with uneventful postoperative recovery.

Our current patient survived infective endocarditis of the AV occurring years prior to presentation. He remains well 7 years following the surgical correction.

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## COMMENTS

### Background

Aortocameral fistulas (ACF) may be congenital or acquired complicating acute aortic dissection following an intimal tear in the vicinity and proximity of the aortic root or after aortic valve (AV) replacement. ACF is an uncommon complication of native AV endocarditis, which is associated with high morbidity and mortality.

### Research frontiers

ACF may originate from any of the three sinuses of Valsalva. Audible continuous murmur may raise suspicion for the presence of ACF. The clinical manifestations of ACF may include exertional dyspnea, chest pain, palpitation, congestive heart failure and recurrent respiratory tract infection. ACF may incidentally be found during routine preoperative examination. Untreated ACF may cause significant morbidity and early mortality.

### Innovations and breakthroughs

The surgical correction of ACF is the treatment of choice but percutaneous transcatheter device intervention has recently been successfully introduced for the closure of ACF. Acquired ACF is an infrequent entity which may occur late after an episode of endocarditis of the native AV.

### Applications

This paper presents a case of acquired aortic-atrial fistulas occurring late after infective endocarditis of the aortic valve, the author reviewed 30 suitable papers and summarized the clinical feature, diagnostic modalities and management of such a disease.

### Peer-review

The authors reviewed the published literature on the aortic-atrial fistulae, but also included several cases of similar connections that occurred between the aorta and other chambers including the ventricles, the left atrium and the pulmonary artery. The interesting side of the manuscript is the review rather than the clinical case. The review is well written and reports a total of 38 cases, presented in different clinical scenario.

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