

# Fatal ascending aorta-to-right ventricle fistula formation after *Staphylococcus aureus* endocarditis of bicuspid aortic valve

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We present a case of a 46-year-old man with advanced acquired immunodeficiency syndrome and congenitally bicuspid aortic valve endocarditis caused by methicillin- and gentamicin-resistant *Staphylococcus aureus*. Endocarditis led to root abscess formation, a complete heart block, and fistulous tract formation between the ascending aorta and the right ventricle. Although perivalvular abscess is not an unusual complication of native valve endocarditis, a fatal fistulous communication between the ascending aorta and the right ventricle is exceedingly rare. (Heart Lung® 2005;34:429–32.)

## CASE REPORT

A 46-year-old man with a history of advanced acquired immunodeficiency syndrome (AIDS) and pancytopenia was admitted for generalized malaise and fever thought to be caused by disseminated *Mycobacterium avium-intracellulare* (MAI) infection.

Physical examination on admission was notable for fever (103°F), tachycardia (120 beats/min), normal blood pressure (120/80 mm Hg), and an absence of heart murmurs. In addition, there were no clinical or radiographic signs of congestive heart failure. Electrocardiogram revealed sinus tachycardia with normal atrioventricular and intraventricular conduction.

On day 2, however, blood cultures drawn on admission grew methicillin-resistant *Staphylococcus aureus* and the patient was started on intravenous vancomycin therapy. A technically difficult transthoracic echocardiogram performed soon afterward revealed thickened aortic valve with mild insufficiency and no significant stenosis, no mobile vegetation on any of the cardiac valves, and normal left and

right ventricular systolic function. Further workup for endocarditis by transesophageal echocardiogram was not pursued at that time.

On day 12, chest pain, shortness of breath, hypoxia, and bradycardia developed in the patient. On physical examination no new murmur was noted.

A new third-degree atrioventricular block with an escape ventricular rhythm in the form of right bundle branch block at 34 beats/min was noted on electrocardiogram and treated with insertion of a temporary transvenous pacemaker.

Because the blood cultures remained positive for methicillin-resistant *S. aureus*, intravenous gentamicin was added to vancomycin therapy. However, within days, gentamicin had to be stopped because the organism was found to be gentamicin resistant.

On day 13, transthoracic and transesophageal echocardiography revealed a mildly to moderately stenotic bicuspid aortic valve with peak and mean gradients of 62 and 31 mm Hg, respectively. Aortic valve area was estimated to be 1.4 cm<sup>2</sup> by continuity equation.

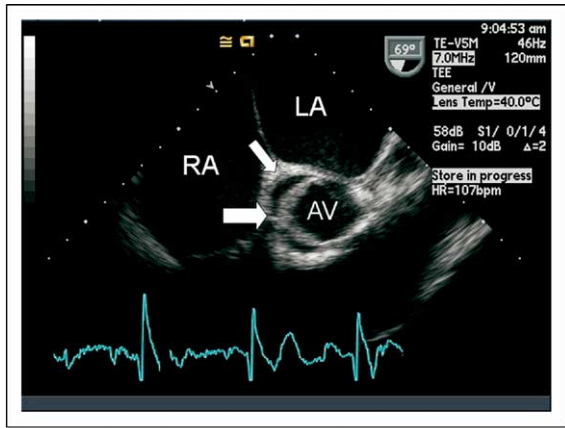
The aortic valve leaflets were unusually thick, and there were at least two mobile echo densities consistent with vegetations (Fig 1). The larger one measured 1.5 cm and was protruding into the ascending aorta, and the smaller one extended into the left ventricular outflow tract.

Furthermore, aortic valve insufficiency was now moderate and had an eccentric jet directed toward the anterior mitral leaflet. The area around the ex-

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**Fig 1** Transesophageal echocardiogram showing bicuspid AV with raphe (*large arrow*). AV leaflets are unusually thickened and studded with vegetations. Periaortic extension of the infection (*small arrow*). AV, aortic valve; RA, right atrium; LA, left atrium.

ternal perimeter of the aortic root was thickened and heaped up, a finding suggestive of periaortic extension of endocarditis.

A cardiothoracic surgery consult was requested for possible aortic valve replacement. However, the patient was deemed inoperable because of his very advanced AIDS and disseminated MAI.

On day 14, oral rifampin at a dose of 600 mg daily was added to vancomycin therapy.

Over the following 7 days the patient was hemodynamically stable with no evidence of congestive heart failure or systemic embolism.

On day 18, the patient became refractory to pacing even with the pacemaker generator set to maximum output amperage. His pulse pressure began to widen, but on auscultation his lungs remained clear.

On day 19, cardiac arrest developed. Transthoracic echocardiogram performed during cardiopulmonary resuscitation (*Fig 2*) revealed that the aortic valve insufficiency remained moderate. However, the following findings were new:

- Continuous color flow from the aortic root into the right ventricle indicative of perforation in the aortic wall, erosion into the right ventricle, and creation of a continuous left-to-right shunt between the ascending aorta and the right ventricle
- Paradoxical movement of the interventricular septum consistent with right ventricular pressure overload
- Severe hypokinesis of the right ventricular free wall

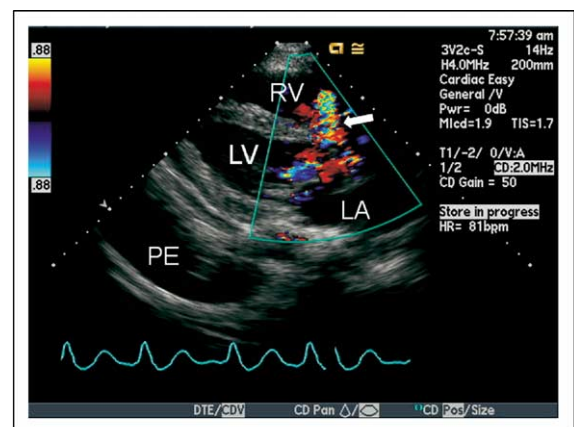
By the end of the transthoracic echocardiography, cardiac standstill developed and the patient was soon pronounced dead.

## DISCUSSION

Perivalvular abscess is a fairly common although under-recognized complication of aortic valve infective endocarditis.<sup>7-9</sup> However, development of an intracardiac fistula after aortic valve infection is a rare occurrence.<sup>10,11</sup>

To date, there is only one other well-documented case of fistulization between the aorta and the right ventricle as a complication of aortic root abscess, although other intracardiac fistulas have been more frequently reported.<sup>1</sup>

Our patient had several risk factors for aortic valve endocarditis. These included a congenitally bicuspid aortic valve, profound immunosuppression caused by advanced AIDS, and pancytopenia caused by disseminated mycobacterial infection. His history of intravenous drug use increased his likelihood of developing a staphylococcal infection, and his history of recurrent hospital admissions increased the likelihood of methicillin resistance. A rather atypical feature of our patient's case was the absence of clinical signs of congestive heart failure until the night before his demise, and even then not because of valvular insufficiency but rather an ascending aorta-to-right ventricular shunt.



**Fig 2** Color Doppler transthoracic echocardiogram revealing a fistulous tract with a left-to-right shunt (*arrow*) from the ascending aorta to the RV as a complication of aortic valve endocarditis. RV, right ventricle; LA, left atrium; LV, left ventricle; PE, left pleural effusion.

Aortic root abscess seems to occur more commonly as a complication of native compared with prosthetic valve endocarditis. In patients with native valve endocarditis the risk is increased when the valve is bicuspid. In a series of 233 patients who developed perivalvular abscess as a complication of infective endocarditis, two thirds of the patients with aortic root abscess had infection of a native valve. One third of the infected valves was bicuspid.<sup>2</sup>

In a retrospective analysis of 25 patients with aortic root abscesses, Lerakis and colleagues<sup>1</sup> observed that 14 (56%) occurred on a native valve, six (43%) of which were bicuspid. Twenty percent developed complete heart block, and four patients developed an intracardiac fistula. One of these was a fistula between the aorta and the right ventricle.

A higher prevalence of aortic root abscess in native valve endocarditis versus prosthetic valve endocarditis was seen in another two series. The percentage of native versus prosthetic aortic valve endocarditis in patients with root abscess was 72% versus 28% in a series of 65 patients<sup>3</sup> and 87% versus 13% in another series of 15 patients.<sup>4</sup>

In the above-mentioned series of 233 patients, significant valvular regurgitation and congestive heart failure were both more commonly associated with native valve endocarditis compared with prosthetic valve endocarditis. High-grade conduction disturbances occurred exclusively in patients with native valve endocarditis. In most other series of patients with perivalvular abscess, New York Heart Association class III or IV heart failure carried a higher mortality and was considered an indication for surgical management.

In a series of 95 patients with infective endocarditis Arnett and Roberts<sup>5</sup> found that all 27 patients with ring abscesses had valvular insufficiency (24 aortic and 3 mitral). They pointed out that classic signs of aortic regurgitation may not be apparent on physical examination in patients with acute aortic insufficiency.

Because of the perivalvular location of the abscess, our patient presented with a new conduction disturbance requiring a permanent pacemaker in the absence of clinical heart failure or hemodynamically significant aortic regurgitation.

Most cases of perivalvular abscess, whether aortic or mitral, are caused by staphylococcal or streptococcal infections. In the previously mentioned series by Lerakis and colleagues,<sup>1</sup> it was found that infection with *S. aureus* carried a much poorer prognosis than infection with other organisms. Compounding this fact in our patient was

methicillin and gentamicin resistance, which limited the spectrum of available antimicrobial therapy.

In our patient, endocarditis developed in the setting of AIDS and pancytopenia caused by disseminated MAI. Infection with the human immunodeficiency virus (HIV)-1 in and of itself does not necessarily increase one's risk for acute infective endocarditis. Nahass et al<sup>6</sup> found that acute infective endocarditis in patients with HIV usually occurred in the setting of intravenous drug use, and that the infection most commonly involved the tricuspid valve. He also found that 75% of these infections were caused by *S. aureus*. In general, the course of endocarditis tends to be the same in asymptomatic patients with HIV-1 infection as in the general population; however, in patients with advanced disease the associated morbidity and mortality are higher.

In conclusion, we presented a case of what initially seemed to be a relatively routine case of endocarditis in a patient with multiple risk factors. However, he developed the less frequently seen complications of aortic root abscess and intracardiac fistula. Given the poor prognosis of perivalvular abscess without surgical treatment, this patient's demise was almost inevitable.

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