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Case Report

Percutaneous transcatheter closure of an aorto-cardiac fistula in a six-year-old Warmblood mare with atrial fibrillation[☆],[☆☆]

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Abstract   This report describes a rare case of an aorto-cardiac fistula in a six-year-old French Warmblood mare presented with atrial fibrillation, decreased performance, ventral oedema, bounding arterial pulsation and pathological jugular venous pulse. A 2.7-cm-diameter fistula connected the right aortic sinus of Valsalva to the right atrium. Atrial fibrillation was likely due to volume overload of the right heart due to left-to-right shunting. The horse was treated by percutaneous transcatheter closure of the fistula delivered under general anaesthesia using a transarterial

approach. The operation was initially successful, and clinical signs of congestive heart failure improved immediately. However, the device dislodged six days after procedure, and the general condition of the horse deteriorated quickly. A second closure attempt to deliver the occluder using a transvenous approach in the standing horse failed, and the horse was eventually euthanized. Procedural aspects and several possible risk factors for device dislodgement are discussed.

Abbreviations

ACF	aorto-cardiac fistula
AF	atrial fibrillation
CHF	congestive heart failure
TTE	transthoracic echocardiography
TOE	transoesophageal echocardiography

A six-year-old, 477 kg French Warmblood mare used for show jumping was referred to the Department of Large Animal Internal Medicine, Ghent University, for evaluation of atrial fibrillation (AF). Six months before presentation, the horse suddenly showed decreased performance associated with excessive sweating and tachypnoea. The mare was only evaluated for musculoskeletal problems, and because no abnormalities were found, she was put at rest for three months. When training was resumed, tachypnoea and sweating during exercise were seen, and another three months of rest was enforced. Only after the six-month rest period was a cardiac arrhythmia detected, and an electrocardiogram at that time indicated AF. The horse was referred for further evaluation.

Examination revealed a pathological jugular venous pulsation, bounding irregular arterial pulse and ventral oedema. Lung auscultation was normal. Cardiac auscultation revealed tachycardia (64 beats/min) with an irregular rhythm typical of AF, a grade 2/6 holosystolic murmur and a grade 2/6 holodiastolic murmur at the left side and a grade 5/6 pansystolic murmur and a grade 5/6 pandiastolic murmur on the right side. Haematology and biochemistry were normal except for a slightly increased gamma-glutamyltransferase concentration of 38 U/L (reference 0—30 U/L). Cardiac troponin I was normal (<0.03 ng/mL). The resting electrocardiogram confirmed AF. Owing to the severity of the clinical signs, electrocardiography during exercise was not performed. Echocardiography[d] showed left and right heart dilation and mild

aortic regurgitation and revealed an aorto-cardiac fistula (ACF) (total length: 29 mm) between the right aortic sinus of Valsalva and right atrium. The diameter of the fistula was 27 mm at the aortic side and 15 mm at the atrial entry, just dorsal to the tricuspid valve. At the atrial side, the fistula showed an irregular, mobile tissue border protruding into the atrium (Fig. 1). Continuous-wave Doppler recordings confirmed the presence of a left-to-right shunt, with a maximal systolic flow velocity of 5.7 m/s, corresponding to a pressure difference of 140 mmHg. Using pulsed wave Doppler, flow reversal towards the heart was detected in the carotid artery during diastole.

It was decided to treat the horse by implantation of an occluder[e] in the ACF. In the month between presentation at the clinic and the implantation, the patient was kept at rest and treated with digoxin (11 µg/kg twice daily PO) and quinapril (0.25 mg/kg twice daily PO). The quinapril therapy was discontinued after four days because the horse became apathetic and showed reduced appetite, possibly due to an additional decrease of the arterial pressure. The procedure itself was initiated in the standing sedated horse. After placement of an 8.5-Fr introducer sheath[f] in the left jugular vein, a pacing catheter[g] was introduced and positioned in the right ventricular apex under echocardiographic guidance. Following induction of general anaesthesia, the ventral portion of the right carotid artery was surgically exposed, a purse-string suture was placed and a 14-Fr introducer sheath[h] was inserted. Subsequently, an angiographic catheter with guidewire[i] was introduced and advanced towards the aortic valve under echocardiographic guidance. The guidewire was inserted in the ACF and
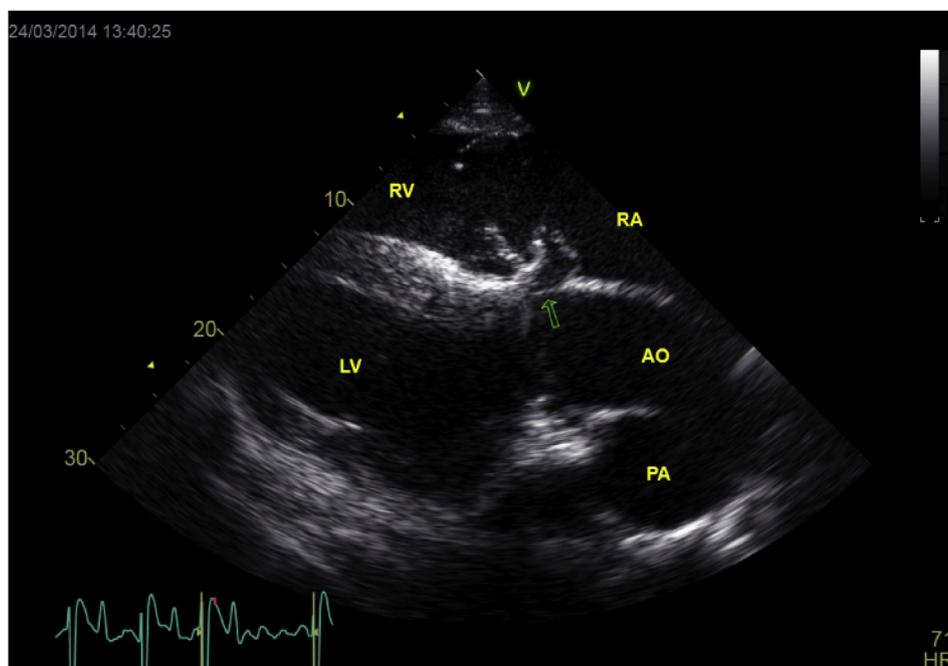

Fig. 1 Echocardiographic image of the aorto-cardiac fistula. Right parasternal left ventricular outflow tract view, demonstrating an aorto-cardiac fistula (green arrow) between the right aortic sinus of Valsalva and the right atrium, just dorsal of the tricuspid valve. At the atrial side, the fistula shows an irregular hyperechoic tissue border that protrudes into the atrium. This was presumed to be fibrous tissue. AO, aorta; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle.

advanced into the right ventricle (Video 1 in Supplementary Material). The angiographic catheter was removed, and the 14-Fr introducer[h] was replaced by a 12-Fr delivery sheath.[j] The delivery sheath was advanced over the wire into the right ventricle (Fig. 2). An occluder,[e] designed to cover a defect size of 15—30 mm, was loaded into the delivery sheath,[j] and the distal disk was deployed in the right atrium. Through gentle traction on the delivery system, the proximal disk was deployed in the aorta. However, even after several attempts, owing to pressure, the proximal disk partially advanced into the fistula (Fig. 3). Before release, device stability was evaluated through gentle pulling and pushing of the device.[e] The implantation resulted in partial closure of the fistula with a minimal residual systolic flow (Fig. 4). The large arterial pressure differences that ensued with fistula closure led to severe reflex bradycardia with transient asystole prompting temporary ventricular pacing at 35 pulses/min. After withdrawal of the 12-Fr delivery sheath,[j] closure of the purse-string suture of the carotid artery and wound closure, a spontaneous supraventricular rhythm

resumed, although it was still irregular due to persistent AF. Recovery was uneventful.

The next day, the horse had an irregular but lower resting heart rate of 35 beats/min, less ventral oedema and a good appetite. The following days, the horse was monitored by clinical examination and echocardiography. The occluder remained in position and did not impede coronary artery flow or valvular function. Appetite was normal, and signs of congestive heart failure (CHF) resolved. However, six days after procedure, the horse was suddenly depressed. Auscultation revealed a heart rate of 40 beats/min with the same heart murmurs as previously described. Echocardiography and thoracic radiography showed dislodgement of the occluder[e] with device migration into the pulmonary artery. Echocardiography identified the occluder in the left pulmonary artery, without apparent obstruction of the pulmonary blood flow.

Supportive treatment consisted of digoxin (11 μg/kg twice daily PO) and intramuscular furosemide (250 mg twice daily); however, the initial clinical signs reappeared within 48 h. Seven days after device embolism, the horse had an increased body temperature (38.7 °C) and signs of laminitis, which resolved with antibiotic (trimethoprim/sulfadiazine) and non-steroidal anti-inflammatory (phenylbutazone) drug treatment.

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[j] Amplatzer™ TorqVue™ 45° delivery system, AGA Medical corporation, Golden Valley, Minnesota, USA.
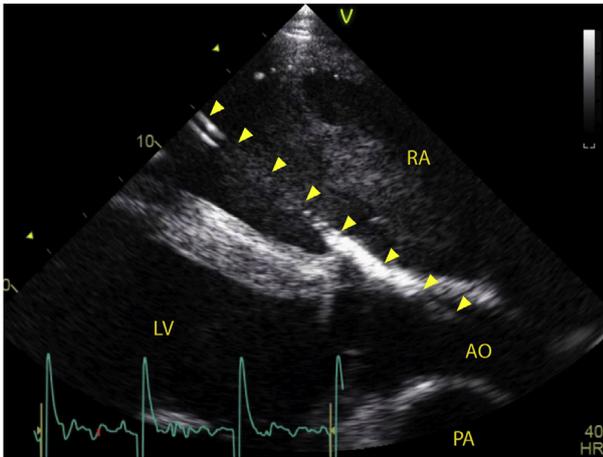
Fig. 2 Echocardiographic image of the delivery sheath. Right parasternal left ventricular outflow tract view, showing the delivery sheath (arrow heads) entering from the aorta through the fistula into the right atrium and entering the right ventricle. AO, aorta; LV, left ventricle; PA, pulmonary artery; RA, right atrium.

Thirteen days after dislodgement, a second attempt to implant an identical occluder[e] was performed in the standing sedated horse, this time using a transvenous approach, anticipating that the slightly larger distal disk might better resist the high pressure of the aorta and remain in place. A 14-Fr introducer sheath[h] was placed in the right jugular vein. Subsequently, a 5-Fr catheter[k] with guidewire[l] was inserted, and it was attempted to position the 0.035″, and subsequently 0.025″, guidewire through the fistula from the right atrium to the aorta. However, owing to cardiac motion, the left-to-right flow and especially the 'wind-sock'-like appearance of the fistulous tract, protruding into the right atrium, the guidewire could not be advanced into the aorta. After repeated attempts, the procedure was aborted and the occluder was not delivered. Three days after procedure, the horse left the clinic with supportive medication (digoxin 11 µg/kg twice daily PO and furosemide 250 mg twice daily IM). Three months later, the horse was euthanized because clinical signs had progressed.

Discussion

Rupture of one of the sinuses of Valsalva, also called 'aortic ring rupture' or 'aortic root rupture',

is an uncommon condition reported in humans [1—10], dogs [11,12] and horses [13—18]. The rupture can communicate with the pericardial space, resulting in cardiac tamponade, or with a cardiac chamber, in which case, the term ACF is used. In horses, ACF is rare and occurs mostly in middle-aged or older stallions, often associated with breeding or exercise [13,18]. Horses can die suddenly or survive for days up to years [13,17,18]. Rupture of a sinus of Valsalva aneurysm is a possible cause of ACF, as well as tearing of the aorta without prior dilation [13,14,16]. Sinus of Valsalva aneurysms can be congenital or acquired. Congenital aneurysms are thought to arise from a deficient tunica media in the area of the sinus due to a separation or incomplete fusion between the annulus fibrosus of the aortic valve and the tunica media of the aorta [13,14,18,19]. The fistula most often extends from the right aortic sinus of Valsalva to the right ventricle and less frequently to the right atrium or the interventricular septum. When the fistula enters the right ventricle or dissects into the interventricular septum, a usually monomorphic ventricular tachycardia is a frequent finding [13,15]. Other common clinical signs include acute colic-like distress, bounding arterial pulses, and signs of CHF [13,15,17]. A continuous murmur loudest over the right side at the level of the fourth intercostal space is typical [13]. In horses, ACF is diagnosed through clinical examination, cardiac auscultation and, especially by, echocardiography [13,14,17,20].

Our case is atypical in several ways. It was a young mare presenting with AF. There was an ACF ending within the right atrium with an associated fibrous tract protruding into the right atrium. Most likely, the ACF occurred six months before admission and explained the exercise intolerance noted at that time. The ACF subsequently led to volume overload and atrial dilation which caused AF to occur. Slowly progressive heart disease can be tolerated for some time; however, superimposition of AF can quickly lead to decompensation with signs of CHF [21]. We speculated that the 'wind-sock'-like appearance at the atrial end of the ACF suggested the lesion started as an aneurysm of the right aortic sinus that eventually ruptured into the right atrium with fibrous tissue formation along the borders. Unfortunately, a necropsy was not obtained to confirm these findings. The mild aortic regurgitation noted could have been caused by the ACF, as this could weaken the aortic ring, or might have been a contributing factor to rupturing of an aortic aneurysm because of secondary aortic dilation [14,16].
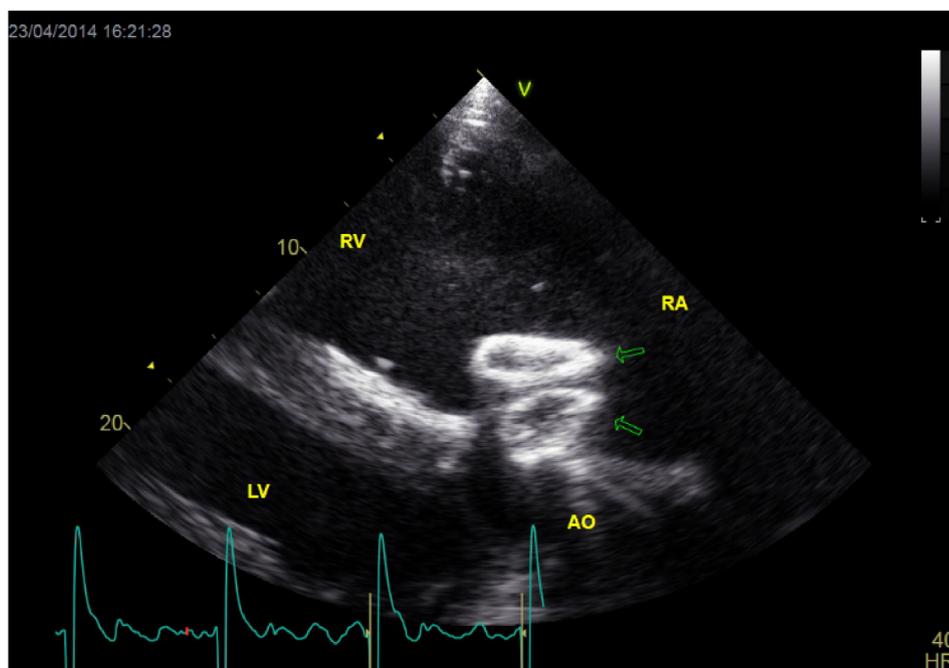
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[k] Amplatz Left 100 cm, N.V. Medtronic Belgium S.A., Brussels, Belgium.
[l] Terumo 260 cm coated straight guidewire 0.035″ and 0.025″, Terumo Europe NV, Leuven, Belgium.

Fig. 3 Echocardiographic view of the implanted occluder. Right parasternal left ventricular outflow tract view, demonstrating the occluder (green arrows) partially located in the fistula. AO, aorta; LV, left ventricle; RA, right atrium; RV, right ventricle.

In humans, ACF is relatively rare. Similarly, the majority of the sinus of Valsalva aneurysms develop in the right and non-coronary aortic sinuses and usually rupture into the right ventricle or atrium [22]. As ACF is difficult to diagnose clinically, transthoracic echocardiography (TTE) and transoesophageal echocardiography (TOE) are used to make a definitive diagnosis [7,8]. In

Fig. 4 Colour Doppler images of the implanted occluder. Right parasternal left ventricular outflow tract view, demonstrating the implanted occluder (green arrows) with minimal systolic flow across the defect. AO, aorta; LV, left ventricle; RA, right atrium; RV, right ventricle.

humans, ACFs are mostly closed through surgical repair, although transcatheter closure has been reported as well. Several devices including ductal occluders, septal occluders, vascular plugs, and coils have been used [1–6,9]. Other occluder types were not applied in our horse but could be considered in future cases. Short-term success rates in humans for transcatheter closure range from 90% to 100% [2,3,5,6] and long-term results seem promising [9]. Complications like device embolization are rare in humans [5,6,9].

Medical treatment is currently the standard treatment for horses with ACF and usually consists of ventricular tachycardia management. This treatment is palliative, as ACF can eventually lead to fatal CHF [13,14,17,18]. Considering the poor prognosis if left untreated, it was decided that the best long-term solution for this mare was transcatheter device implantation. The device remained stable for five days during which the horse's clinical signs of CHF resolved, suggesting successful defect correction. There is one report of successful transcatheter closure of an aorto-right ventricular fistula in a transarterial approach in an 18-year-old Thoroughbred stallion [16]. Probable reasons why the occluder stayed in position in that case are the smaller defect size and the absence of a fibrous tunnel-like protrusion interfering with device deployment.
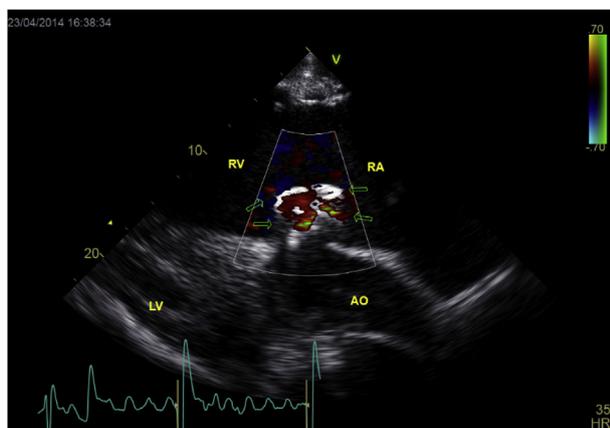
In humans, fistula dimension assessment and device implantation are performed under fluoroscopic and TOE or TTE guidance [2,3,5—7,9,10,23]. Device selection is based on the largest diameter of the aortic end of the defect as this is the largest part of the defect [2,5]. However, some authors based device selection on the narrowest diameter of the ACF [3,6,9]. This is also advised in the instruction manual of the Amplatzer™ Duct Occluder, the device most commonly used for ACFs in humans [24]. Using either measurement protocols, a device 1—5 mm larger in diameter is chosen because of the frail borders of the defect [2,3,5,6,9,23]. The device chosen in this case was an Occlutech Figulla® Flex Atrial Septal Defect occluder suitable for defects ranging 15—30 mm diameter, which sufficiently covers the defect diameter of 27 mm. Using a larger occluder might have resulted in a more stable position but could also have increased the risk for hampering coronary artery flow or aortic valve function.

In the only report of transcatheter occlusion of an ACF in a horse, defect sizing and device implantation were accomplished with TTE guidance [16], as in the present case. Fluoroscopy or TOE is not applicable in horses due to their long neck and voluminous thorax, although Young et al. (1995) performed TOE with a custom-made probe in anaesthetized horses for experimental purposes [25].

The first attempt to position the occluder was a transarterial approach with the occluder advanced from the carotid artery into the aorta. This facilitated positioning of the device because it was advanced in the same direction as the shunt flow. However, ASD occluders are designed for a transvenous approach. The distal retention disk, which is deployed first, has the largest diameter and should ideally be positioned on the higher pressure (aortic) side. The smaller proximal disk is designed to be positioned on the right atrial side of an atrial septal defect [26]. In this case, the smaller retention disk was deployed on the aortic side where it was exposed to higher pressures than it was designed for. This and the location of the disc within the fistula itself were not ideal for maintaining the occluder's position. We later tried a transvenous approach. This technique avoided general anaesthesia and should be beneficial for occluder fixation. However, this approach was technically more difficult, and we were unable to advance the guidewire into the fistula. In people, a procedure is described for a transvenous approach in which an arteriovenous loop is created to advance the occluder against the strong left-to-right shunt [2,4]. Further experience is needed to see if this technique and available guidewires might be applicable in horses.

A second and probably more important cause for the occluder dislodgement was the anatomy of the fistulous tract. Based on TTE (Fig. 3), the device was located within the fistula, making the waist of the occluder very long. This was probably caused by the relatively rigid border and length of the fistula and this prevented full device deployment, so the device could not effectively and stably occlude the defect. Retrieval of the embolized occluder [27] was not attempted. The length of commercial retrieval devices would probably have been borderline, and TTE visualisation was thought to be insufficient for procedure guidance.

In conclusion, transcatheter occlusion using ultrasound guidance is a feasible technique to consider for closure of an ACF entering the right atrium. Defect size, length, and morphology must be considered for successful device deployment. Further experience is needed to refine the procedures and to define the risk factors for occluder dislodgment in horses with ACF.

Conflict of Interest Statement

The authors do not have any conflicts of interest to disclose.

Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jvc.2019.06.003.

Supplementary Material

| 1 | Intraprocedural video of the guidewire advancement. | This right parasternal left ventricular outflow tract view shows how the guidewire was manoeuvred from the aorta through the aorto-cardiac fistula into the right atrium. AO, aorta; LV, left ventricle; PA, pulmonary artery; RA, right atrium. |

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