Double Outlet Right Atrium in an American Domestic Shorthair Cat

Matthew Denton, DVM, Jordan P. Vitt, DVM, DACVIM (Cardiology), Ashley B. Saunders, DVM, DACVIM (Cardiology), and Sonya Wesselowski, DVM, MS, DACVIM (Cardiology), *College Station, Texas*

INTRODUCTION

Double outlet right atrium (DORA) is a type of atrioventricular septal defect (AVSD) that has been rarely reported in the domestic cat (*Felis catus*).[1-3] One prevalence study of congenital heart disease in cats reported 6 AVSDs out of the 57,418 cats whose records were evaluated, none of which had DORA morphology.[1] Another study describing the natural history of AVSDs in 26 cats reported that only 4 of the cases had DORA morphology.[2] Clinical presentation is variable, with clinical signs ranging from vomiting and hyporexia to respiratory distress.[2,3] Point-of-care ultrasound in the emergent setting can guide treatment and prompt pursuit of transthoracic echocardiography (TTE) to diagnose this congenital cardiac abnormality.[4] Here we describe the presentation, diagnostic findings, and treatment of a rare case of DORA in a domestic cat with a focus on TTE imaging.

CASE PRESENTATION

A 3.5-year-old male castrated American domestic shorthair cat was presented emergently for a 3-day history of vomiting, hyporexia, and intermittent increased respiratory effort. Prior to presentation, same day referral thoracic radiographs were obtained (Figure 1) and revealed marked cardiomegaly with biatrial enlargement and a globoid cardiac silhouette suggestive of pericardial effusion. Distension of both pulmonary arteries and pulmonary veins was appreciable, as was a small volume of pleural effusion and a diffuse, mild interstitial pattern with peribronchial cuffing. Radiographic findings were consistent with congestive heart failure (CHF). The prioritized differential list at that time included hypertrophic cardiomyopathy, congenital cardiac disease such as mitral valve dysplasia, cor triatriatum sinister (CTS), or other complex congenital abnormalities. Physical examination revealed increased bronchovesicular sounds, tachypnea, open mouth breathing, tachycardia with an irregular rhythm, and a grade II/VI left parasternal systolic heart murmur. Point-of-care ultrasound revealed a markedly enlarged left atrium (LA), gall bladder wall edema, and a distended caudal vena cava.

From the Department of Small Animal Clinical Sciences, School of Veterinary Medicine and Biomedical Sciences, Texas A&M University, College Station, Texas.

Keywords: Feline, Veterinary, Congenital, Echocardiography

Correspondence: Sonya Wesselowski, DVM, MS, DACVIM (Cardiology), Department of Small Animal Clinical Sciences, 4474 TAMU, College Station, TX 77843. (E-mail: *swesselowski@cvm.tamu.edu*).

2468-6441

https://doi.org/10.1016/j.case.2023.12.026

VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE, right parasternal 4-chamber long-axis view, demonstrates leftward deviation of the apical IAS creating a division of the markedly dilated LA into a proximal and distal chamber due to DORA morphology. The left and right AV valves insert at the same level consistent with an AVSD. There is mild pericardial effusion between the pericardium and LV free wall and the LA.

Video 2: Two-dimensional TTE, right parasternal 4-chamber long-axis view with simultaneous color-flow Doppler, demonstrates continuous, turbulent flow from the proximal to distal LA across the obstruction created by the leftward deviated apical IAS due to DORA.

Video 3: Two-dimensional TTE, right parasternal long-axis 4-chamber view, during an agitated saline contrast study performed via right cephalic vein injection, demonstrates the sequential appearance of intracardiac microbubbles from the RA to the distal LA chambers and then the simultaneous entrance into both the LV and RV. There are no microbubbles noted in the proximal LA chamber.

View the video content online at www.cvcasejournal.com.

Transthoracic echocardiography was then pursued, which confirmed the marked LA and left atrial appendage (LAA) dilation with spontaneous echogenic contrast appreciated within the LAA. The LA to aortic root ratio was 4.1 (normal reference range, 1.0-1.4).[5] The ventricles were oriented in d-loop morphology. The left and right atrioventricular (AV) valves inserted at the same level, consistent with absence of the AV septum. An ostium primum atrial septal defect (ASD) was present, along with a marked leftward deviation of the apical interatrial septum (IAS; Figure 2, Video 1). An inlet ventricular septal defect (VSD) was also identified on two-dimensional imaging; however, no flow was detected across this region with Doppler imaging due to adherence of AV valve tissue to the crest of the interventricular septum (Figure 3). A single AV ring with 2 AV orifices was appreciated, along with a cleft anterior left AV valve leaflet (Figure 3). The deviated IAS resulted in a marked obstruction to blood flow from the proximal to distal LA, with continuous high-velocity flow (peak, 2.25 m/sec; estimated gradient, 20 mm Hg) across the obstruction (Figure 4, Video 2). Measurement of right atrial (RA) diameter as an assessment of size was challenging due to the abnormal position of the IAS; however, the chamber was subjectively considered to be severely enlarged. Right ventricular (RV) chamber dilation based on an RV internal diameter at end diastole of 1.34 cm (compared to previously published control population data with a

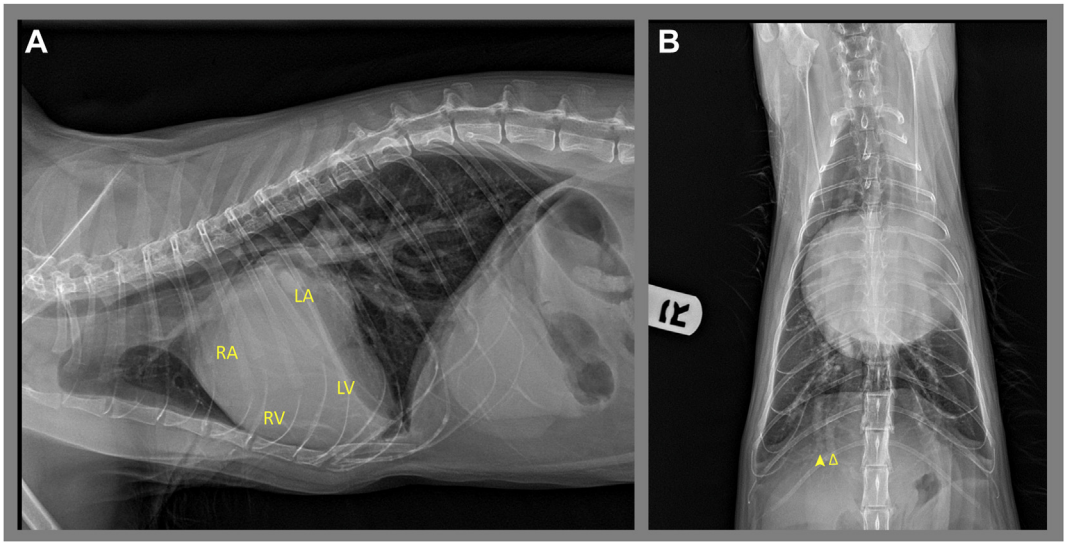

Figure 1 Thoracic radiograph findings in lateral **(A)** and dorsoventral **(B)** demonstrate globoid cardiomegaly, biatrial enlargement, dilated pulmonary arteries (*arrowhead*) and veins (*open arrowhead*), a small pleural effusion, and mild interstitial pattern with peribronchial cuffing.

mean ± SD of 6.7 ± 1.4 mm)[6] and concentric hypertrophy of the RV free wall based on an RV free wall thickness at end diastole of 4.4 mm (compared to previously published control population data with a mean ± SD of 2.4 ± 0.4)[6] were also appreciated. Valvular regurgitation of both the right and left AV valves was identified, with the velocity of the right AV valve regurgitation suggestive of pulmonary hypertension with an RV pressure estimate of at least 45 mm Hg. There was intermittent echogenic dropout of the IAS in the location of the fossa ovalis but no color or spectral Doppler flow suggestive of patency across the area. Mild pericardial effusion was noted as well, suspected to be secondary to CHF. A saline contrast study was performed by injecting 2 mL of agitated saline into the right cephalic vein. Intracardiac microbubbles were seen entering the right atrium (RA) and the distal LA chamber simultaneously and then exiting into both the left ventricle (LV) and right ventricle (RV); (Figure 5, Video 3). No microbubbles entered the proximal LA. The TTE findings were consistent with a diagnosis of DORA, subclassified as a DORA secondary to atrial septal malalignment in conjunction with a transitional AVSD.[2,7-9] Based on the TTE findings, the source of the heart murmur was likely related to a combination of turbulent blood flow between the proximal and distal LA chambers and AV valve regurgitation.

Contemporaneous electrocardiogram (ECG) during the TTE was consistent with atrial fibrillation with a rapid ventricular response rate ranging between 220 and 300 beats per minute. A 6-lead ECG confirmed the diagnosis of atrial fibrillation conducted with right bundle branch block (RBBB; Figure 6).

The presumed pathophysiology of the cat's congenital lesion and resultant decompensation into overt CHF began with obstruction to blood flow between the proximal and distal LA chambers leading to elevated pressure within the proximal LA and dilation of this chamber. Over time, increased LA pressure led to postcapillary pulmonary hypertension and subsequent right-sided cardiac remodeling including RV hypertrophy and RA dilation. The marked atrial enlargement predisposed the cat to developing atrial fibrillation, with the onset of this tachyarrhythmia likely precipitating the clinical decompensation into CHF that manifested as pulmonary edema and small-volume pleural and pericardial effusion.

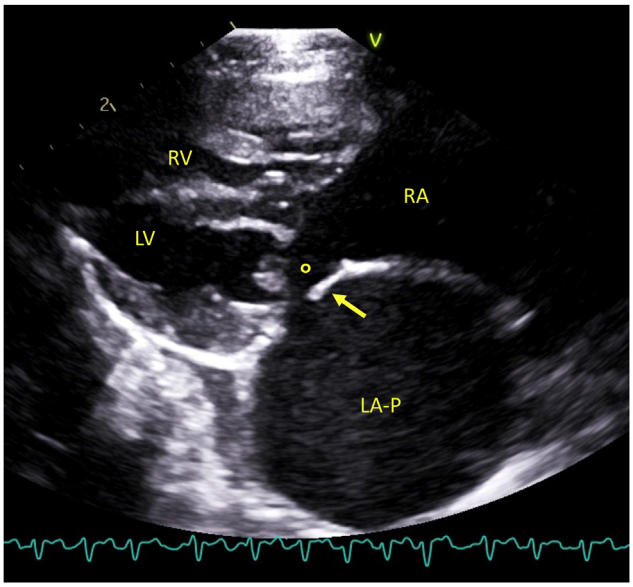

Figure 2 Two-dimensional TTE, right parasternal 4-chamber long-axis view in early systole, demonstrates leftward deviation of the apical IAS (*arrow*) resulting in division of the LA into a dilated proximal chamber (LA-P) and a distal chamber (°). The left and right AV valves insert at the same level (AVSD), and the dilated RA communicates with both the RV and the LV via the distal LA chamber (DORA). The RV free wall is hypertrophied, and the ECG demonstrates atrial fibrillation. *LA-P*, Proximal left atrial chamber.

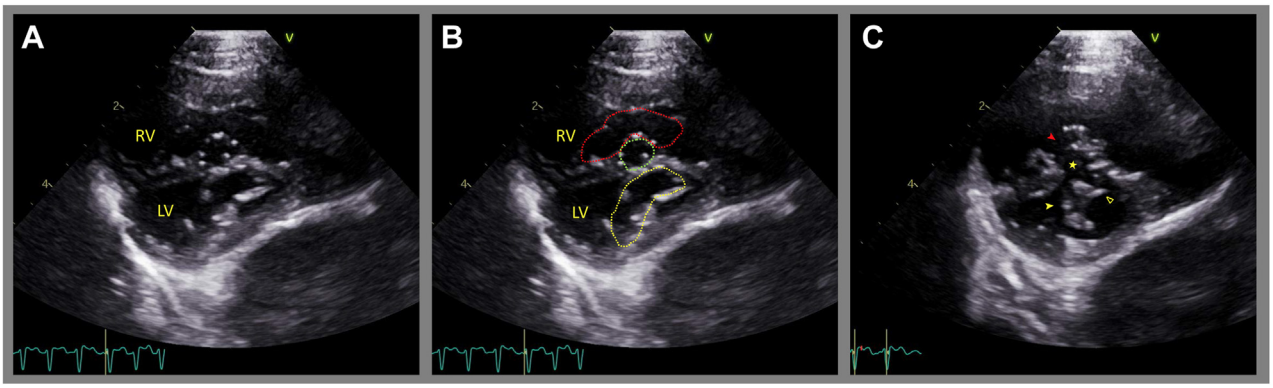

Figure 3 Two-dimensional TTE, right parasternal basal short-axis diastolic view without **(A)** and with **(B)** distinct left and right AV valve orifice tracings, demonstrate a ring of AV valve tissue overriding a region of deficient muscular interventricular septal tissue (inlet VSD). The right AV valve (*red line*), left AV valve (*yellow line*), and ring of AV valve tissue between the 2 (*green line*) are shown. **(C)** In midsystole, the right AV valve (*red arrowhead*), left AV valve (*yellow arrowhead*), a cleft in the anterior left AV valve (*open yellow triangle*), and the region of the muscular interventricular septum with deficient tissue (*yellow star*) are shown.

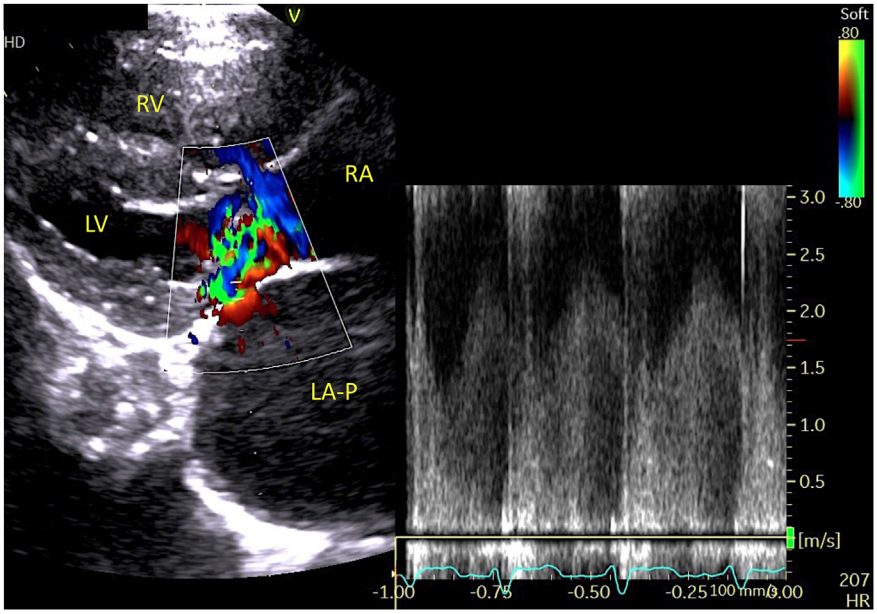

Figure 4 Two-dimensional TTE with color flow–guided pulsed-wave Doppler, right parasternal 4-chamber long-axis systolic view, demonstrates continuous, turbulent flow from the proximal to distal LA across the obstruction created by the DORA. The minimum and maximum velocities are 1.5 m/sec and 2.25 m/sec, respectively. *LA-P*, Proximal left atrial chamber.

Due to poor prognosis, the cat was discharged at the owner's request for palliative at-home care. Medical therapy consisted of 5 mg furosemide (1.1 mg/kg) by mouth twice daily, 15 mg diltiazem HCl XR (3.2 mg/kg) by mouth twice daily, 18.75 mg clopidogrel bisulfate by mouth once daily (4 mg/kg), 8 mg maropitant citrate by mouth once daily (1.7 mg/kg), and mirtazapine 2% topical ointment to be applied once daily to the hairless pinnae. The patient continued to clinically decline at home and was re-presented 3 days later for humane euthanasia. A necropsy was declined.

DISCUSSION

Double outlet right atrium is a congenital cardiac defect that is rare and not well described in domestic cats.[1-3] It is a type of AVSD that allows the RA to empty simultaneously into both the LV and RV.[10] Embryologically, the development of DORA can relate to malalignment of the IAS or malalignment of the interventricular septum.[9] Appropriate alignment of the septa is dependent on normal endocardial cushion development.[11] In patients with DORA, it has been proposed that the embryologic septum primum deviates leftward and partially fuses in an abnormal fashion with the left lateral endocardial cushion rather than fusing at the crux of the heart.[8-10] In this scenario, the LA is divided into a proximal and distal chamber and communication between the RA and distal LA exists via an ostium primum ASD. If, however, the embryologic septum primum deviates so far leftward that it completely fuses with the left lateral endocardial cushion, left AV valve atresia can result. In this second scenario, communication between the right and left atria can only occur via the fossa ovalis.

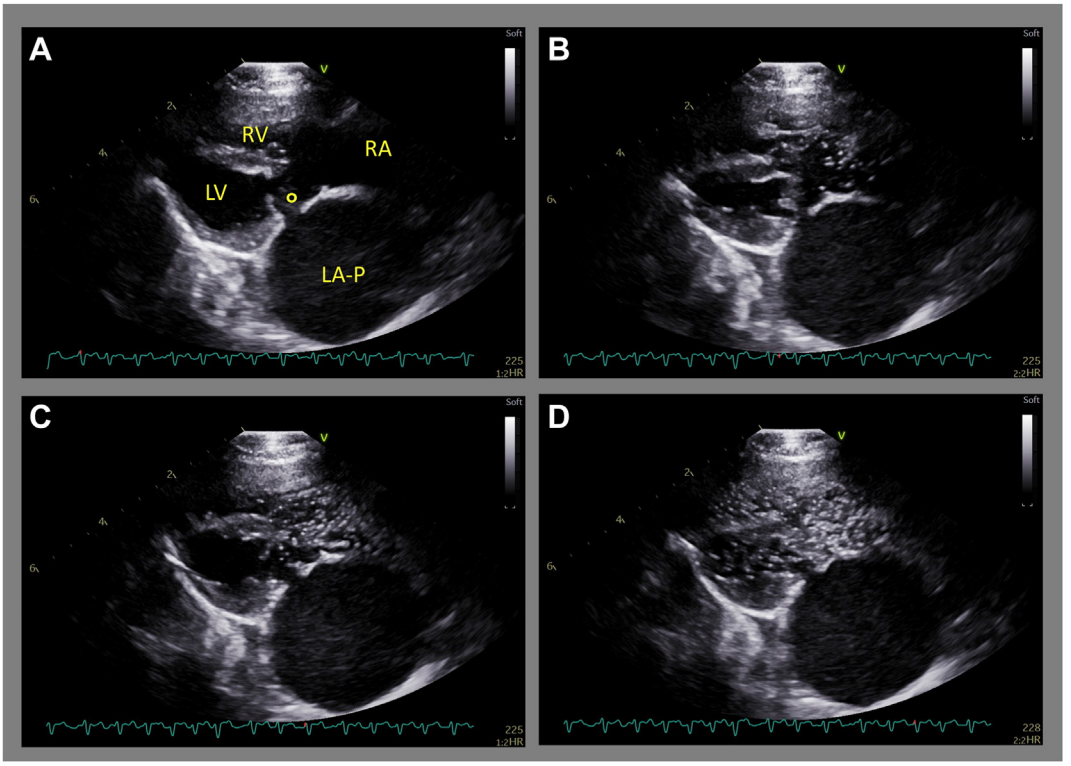

Figure 5 Two-dimensional TTE, right parasternal long-axis 4-chamber view, during an agitated saline contrast study performed via right cephalic vein injection, demonstrates baseline **(A)**, appearance of intracardiac microbubbles in the upper and mid RA **(B)**, then the lower RA and distal LA chambers (°; **C**), and then simultaneous entrance of microbubbles into both the LV and RV **(D)** with no microbubbles ever noted in the proximal LA. *LA-P*, Proximal left atrial chamber.

Additionally, the right AV valve straddles the muscular ventricular septum over a VSD, allowing for ejection of blood into both the RV and LV, although the LV is often hypoplastic in this scenario. While this represents another form of DORA,[7,12] it is anatomically distinct from the first scenario described. The cat of the present case was consistent with the former DORA scenario in combination with a transitional AVSD. An ostium primum ASD and a VSD were present; however, flow through the VSD was absent due to AV valve tissue adhering to the crest of the muscular interventricular septum, as previously described by Gupta *et al.*[7]

In patients with a divided LA, differential diagnoses other than DORA must also be considered, including CTS and a supramitral ring. Both of the latter conditions result in a proximal and distal LA; however, the dividing membrane in a CTS is above the LAA, while the dividing membrane is beneath the LAA with a supramitral ring. In the setting of DORA with an ostium primum ASD (as appreciated in this case), the LAA is above the dividing membrane (similar to a supramitral stenosis); however, the dividing membrane is able to be recognized as a leftward deviated IAS. In CTS and supramitral ring cases, the IAS is expected to be normally positioned in the absence of concurrent congenital abnormalities. Additionally, DORA is a subtype of AVSD, thus absence of the AV septum with the left and right AV valves inserting at the same level is expected in cases with DORA but not in cases with solely CTS or a supramitral ring. Finally, contrast echocardiography using agitated saline can be used to help confirm DORA by observing intracardiac microbubbles moving from the RA into the distal LA and then simultaneously into both ventricles.

Conduction system abnormalities are well recognized in humans with AVSDs.[13] Similar conduction system abnormalities have been reported in the limited number of described cats with AVSDs.[2,3] More specifically, 4 out of 5 of the previously reported feline DORA cases were concurrently diagnosed with partial or complete RBBB.[2,3] This was also noted in the cat of this report, suggesting that identification of partial or complete RBBB in cats should prompt clinicians to consider AVSDs and DORA as possible differential diagnoses. In the cat of the present case, in addition to a conduction disturbance, atrial fibrillation also developed due to the marked atrial dilation.

Despite the hemodynamic consequences of DORA, cats with this congenital defect can remain asymptomatic into adulthood, as demonstrated by the cat of this report, who was 3.5 years old at the time of diagnosis. Of the 5 previously reported feline DORA cases in the literature, 3 of 5 were adult cats at the time of their diagnosis, with ages of 5.1, 9.0, and 12.9 years.[2,3] The other 2 reported cats were juvenile when diagnosed (5 months and 11 months).[2] The majority of cats with DORA described in the literature presented with CHF, as did the cat of this report, thus prompting echocardiographic examination and subsequent diagnosis of the defect. The reason that some cats remain asymptomatic into adulthood is not entirely clear, although superior LA compliance in some cats versus others likely plays some role. In human patients with CTS that do not become symptomatic until adulthood it has been postulated that clinical decompensation may relate to the onset of fibrosis or calcification of the dividing atrial membrane, development of mitral regurgitation over time, or the onset of atrial fibrillation.[14] These factors could also

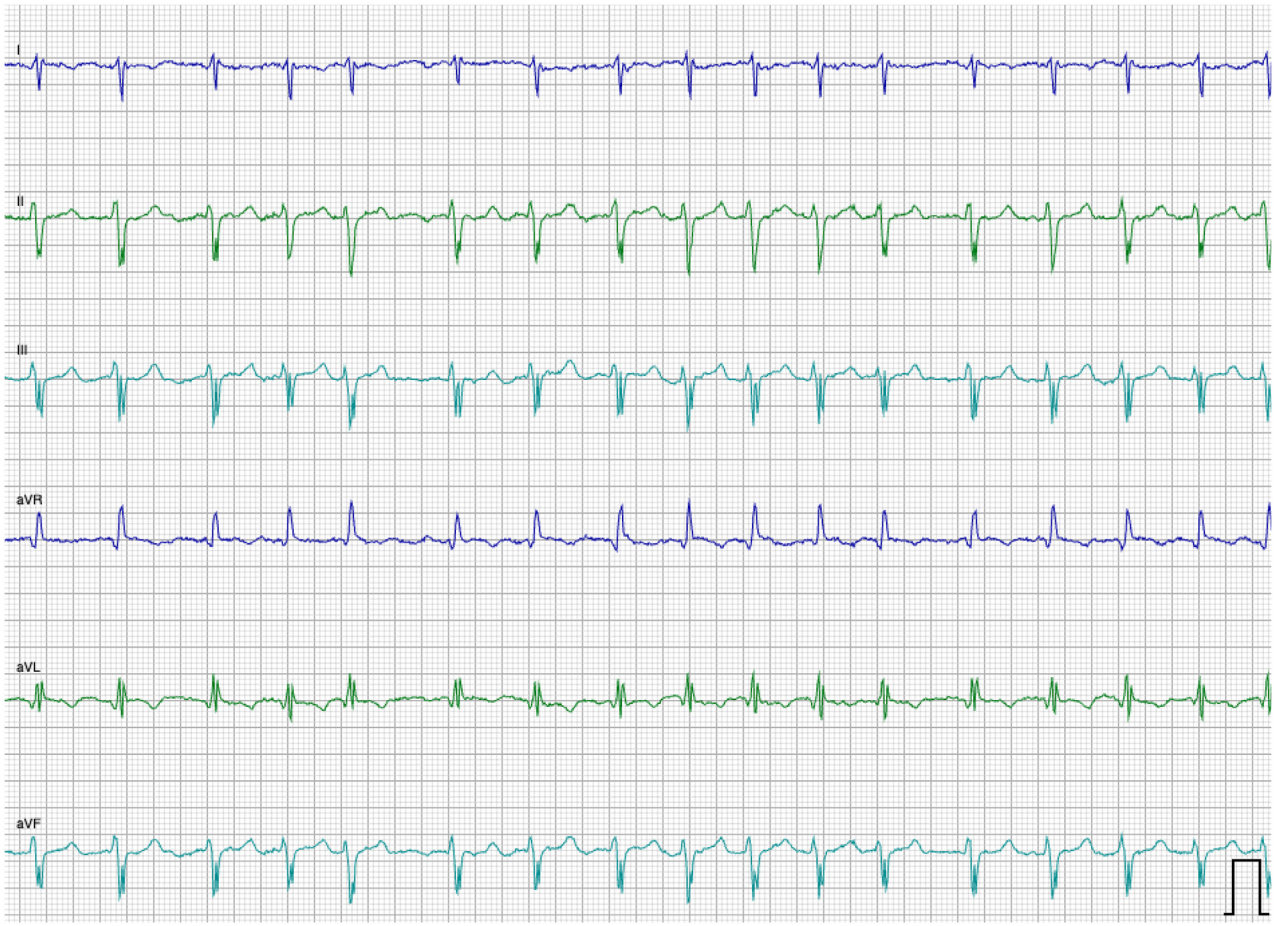

Figure 6 Six-lead surface ECG performed in right lateral recumbency. Calibration: 50 mm/second, 10 mm/mV. Note the absence of P waves, irregular ventricular response rate, and wide, negative QRS complexes (60 msec) consistent with atrial fibrillation and a RBBB pattern.[22] The mean heart rate was 220 beats/minute, and the electrical axis was approximately −120°.

play a role in cats with DORA. In the cat of the present case report, the development of atrial fibrillation is thought to have played a role in triggering clinical decompensation.

In humans the treatment of choice for DORA is surgical correction under cardiopulmonary bypass.[9,10,15] Cardiopulmonary bypass remains largely out of reach for client-owned cats due to technical challenges associated with small body size and expense.[16] While bypass has been successfully performed in the cat in a research setting, studied cats were systemically healthy, had no cardiovascular disease, and were euthanized under anesthesia after being weaned from bypass.[17] A singular case report exists of a client-owned cat undergoing bypass successfully for surgical repair of an ostium secundum ASD.[18] If cardiopulmonary bypass and open-heart surgery become more feasible in cats in the future, advanced imaging studies such as ECG-gated computed tomography angiography and cardiac magnetic resonance imaging would be ideal for surgical planning. Until then, the risks of advanced imaging in cats may outweigh the benefits due to the need for general anesthesia.

The default treatment for cats with DORA at this time is medical management consisting of diuretic therapy, antithrombotic medications, and antiarrhythmic therapy, as needed. Improved echocardiographic surveillance of young cats with heart murmurs may identify cases of DORA before they progress into CHF. If cats with DORA are identified in a preclinical stage, client education regarding the early signs of CHF and monitoring of resting respiratory rate at home may allow for earlier recognition of CHF when it eventually occurs. This may prevent emergency room presentation and hospitalization by recognition of CHF in the earliest stages and initiation of diuretic therapy on an out-patient basis. Additionally, awareness of underlying heart disease in the preclinical stage allows for avoidance of unnecessary anesthetic events, elective intravenous fluid administration, and long-acting glucocorticoid administration, all of which could prompt clinical decompensation.[19] Finally, although preclinical medical management of cardiac disease in cats is often limited, administration of antiplatelet medications can be considered when LA enlargement is identified on TTE to decrease the likelihood of thromboembolic events.[20] Once CHF has developed, minimally invasive procedures such as atrial septostomy of the deviated IAS could potentially be considered as a palliative treatment option, as it may delay the progression of CHF as it has in other types of cardiac disease, resulting in elevated LA pressure in veterinary medicine.[21]

CONCLUSION

Double outlet right atrium is a rare congenital heart defect in the cat that can be diagnosed using TTE. Despite the hemodynamic consequences of DORA, cats with this defect can remain undiagnosed until adulthood.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with the following guidelines (detailed in box below): The work was descriptive only and written retrospectively. At the author's institution, this does not require Institutional Animal Care and Use Committee approval.

CONSENT STATEMENT

The authors declare that since this was a noninterventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status. This was a case report of a veterinary patient (cat) written retrospectively and was descriptive only.

FUNDING STATEMENT

The authors declare that this report did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

DISCLOSURE STATEMENT

The authors report no conflict of interest.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2023.12.026.

REFERENCES

1. Schrope DP. Prevalence of congenital heart disease in 76,301 mixed-breed dogs and 57,025 mixed-breed cats. J Vet Cardiol 2015;17:192-202.
2. Schrope DP. Atrioventricular septal defects: natural history, echocardiographic, electrocardiographic, and radiographic findings in 26 cats. J Vet Cardiol 2013;15:233-42.
3. Durham J, Maisenbacher H. Double-outlet right atrium in a 9 year-old cat. J Vet Cardiol 2014;16:127-31.
4. Ward JL, DeFrancesco TC. The role of point-of-care ultrasound in managing cardiac emergencies. Vet Clin North Am Small Anim Pract 2023;53:1429-43.
5. Abbott JA, MacLean HN. Two-dimensional echocardiographic assessment of the feline left atrium. J Vet Intern Med 2006;20:111-9.
6. Visser LC, Sloan CQ, Stern JA. Echocardiographic assessment of right ventricular size and function in cats with hypertrophic cardiomyopathy. J Vet Intern Med 2017;31:668-77.
7. Gupta SK, Gupta A, Ramakrishnan S, Anderson RH. Clarifying the atrioventricular junctional anatomy in the setting of double outlet right atrium. Ann Pediatr Cardiol 2015;8:233-9.
8. Scansen BA, Schneider M, Bonagura JD. Sequential segmental classification of feline congenital heart disease. J Vet Cardiol 2015;17(Suppl 1):S10-52.
9. Brancaccio G, Amodeo A, Rinelli G, Filippelli S, Sanders SP, Di Donato RM. Double-outlet right atrium: anatomic and clinical considerations. Ann Thorac Surg 2007;83:619-21.
10. Van Mierop LHS. Pathology and pathogenesis of endocardial cushion defects. Surgical implications. In: Second Henry Ford Hospital International Symposium on Cardiac Surgery. New York: Appleton-Century-Crofts; 1977:201-7.
11. Cooper RS. Endocardial cushion defects: embryology, anatomy and pathophysiology. Adv Cardiol 2004;41:118-26.
12. Shetkar SS, Kothari SS. Double-outlet left atrium: ventriculo-atrial malalignment defect. Ann Pediatr Cardiol 2013;6:158-61.
13. Feldt RH, DuShane JW, Titus JL. The atrioventricular conduction system in persistent common atrioventricular canal defect: correlations with electrocardiogram. Circulation 1970;42:437-44.
14. Alphonso N, Nørgaard MA, Newcomb A, d'Udekem Y, Brizard CP, Cochrane A. Cor triatriatum: presentation, diagnosis and long-term surgical results. Ann Thorac Surg 2005;80:1666-71.
15. Ambra M, Nuri HA, Giuseppe P, Marasini M, Francesco S. Double-outlet right atrium: review of a rare anomaly with an exemplary case [published correction appears in World J Pediatr Congenit Heart Surg. 2020 Jul;11(4):391]. World J Pediatr Congenit Heart Surg 2020;11:79-84.
16. Pelosi A, Anderson LK, Paugh J, Robinson S, Eyster GE. Challenges of cardiopulmonary bypass-a review of the veterinary literature. Vet Surg 2013;42:119-36.
17. Brourman JD, Schertel ER, Holt DW, Olshove VA. Cardiopulmonary bypass in the cat. Vet Surg 2002;31:412-7.
18. Uechi M, Harada K, Mizukoshi T, Mizuno T, Mizuno M, Ebisawa T, et al. Surgical closure of an atrial septal defect using cardiopulmonary bypass in a cat. Vet Surg 2011;40:413-7.
19. Côté E. Feline congestive heart failure: current diagnosis and management. Vet Clin North Am Small Anim Pract 2017;47:1055-64.
20. Hogan DF, Fox PR, Jacob K, Keene B, Laste NJ, Rosenthal S, et al. Secondary prevention of cardiogenic arterial thromboembolism in the cat: the double-blind, randomized, positive-controlled feline arterial thromboembolism; clopidogrel versus aspirin trial (FAT CAT). J Vet Cardiol 2015;17(Suppl 1):S306-17.
21. Allen JW, Phipps KL, Llamas AA, Barrett KA. Left atrial decompression as a palliative minimally invasive treatment for congestive heart failure caused by myxomatous mitral valve disease in dogs: 17 cases (2018-2019). J Am Vet Med Assoc 2021;258:638-47.
22. Santilli R, Moïse SN, Pariaut R, Perego M. Electrocardiography of the Dog and Cat: Diagnosis of Arrhythmias. Edra S.p.A.; 2018.