Double-Outlet Right Atrium in a Young Cat

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INTRODUCTION

Double-outlet right atrium (DORA) is a rare congenital heart defect characterized by a right atrium (RA) that empties into both the right ventricle (RV) and the left ventricle (LV) due to either leftward deviation of the atrial septum, which fuses to the left atrium (LA) free wall, causing various degrees of obstruction to the LA outflow, or to rightward deviation of the interventricular septum. This anomaly is a type of endocardial cushion defect and has been reported in humans. However, only sparse information about this malformation is available in veterinary medicine. This case report describes the clinical signs and diagnostic findings of a young cat diagnosed with DORA.

CASE PRESENTATION

A 9-month-old female neutered domestic shorthair cat (3.5 kg body weight) was referred to the Small Animal Teaching Hospital at the University of Liverpool for investigation of acute-onset tachypnea and dyspnea. These clinical signs were first noted 2 days prior to admission. Thoracic radiographs performed by the referring veterinary surgeon revealed a generalized interstitial pulmonary pattern.

On presentation, marked dyspnea and tachypnea (80 breaths/min) were noted. Cardiac auscultation revealed a grade II/VI left parasternal systolic murmur and a grade I/VI right-sided systolic murmur. The heart rate was 200 bpm with a regular rhythm and fair quality peripheral pulses. The remainder of the clinical examination was unremarkable.

The cat received oxygen supplementation and intravenous furosemide boluses to a total dose of 6 mg/kg over 12 hours, resulting in a significant improvement of the breathing rate and effort.

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Venous blood gas analysis showed no significant abnormalities, and systolic blood pressure, measured via Doppler technique, was 150 mm Hg. A 6-lead electrocardiogram demonstrated the presence of a sinus tachycardia with heart rate of 220 bpm.

A transthoracic echocardiogram (TTE) with Doppler was performed. The interatrial septum (IAS) was deviated leftward, and its distal end was noted to be thickened, globular, and connected with the apical border of the LA's free wall (Figure 1, Video 1), resulting in obstruction of the LA outflow. The LA outflow traversed a small ostium primum atrial septal defect (ASD), shown by the presence of a high-velocity, diastolic flow across this obstruction evident on both color-flow and spectral Doppler (Figures 2 and 3, Video 2). As a result of the IAS deviation, the dilated LA was not in direct communication with the mitral valve apparatus. Distended pulmonary veins entered the dilated LA. The LA was in connection with a normally located, markedly dilated left atrial appendage (LAA) and with the extension of the RA ventrally. The RA was in turn connected with the larger morphological LA medially and dorsally and in direct communication with both tricuspid and mitral valve apparatus ventrally (Figures 1 and 2, Videos 1 and 2).

The aorta and pulmonary arteries were normally located, and there was no evident conotruncal malformation. There was laminar aortic and pulmonic flow at expected velocities. No ventricular septal defect was observed. The atrioventricular (AV) valves were both located at a similar plane without obvious apical displacement of the tricuspid valve. Subjectively, the mitral valve septal leaflet appeared elongated, and on the short-axis view this valve had a trifoliate structure with the zone of apposition between the bridging leaflets directed toward the interventricular septum. This was associated with moderate systolic mitral regurgitation directed into the RA (Videos 3 and 4).

Bidirectional shunting was observed on color Doppler assessment. Left-to-right shunting was noted as the LA, and the LV delivered blood into the extension of the RA, via the ASD and the mitral valve regurgitation, respectively. Right-to-left shunting was observed as the RA, being in direct communication with the mitral valve apparatus, drained into the LV. This bidirectional shunting resulted in mixing of oxygenated and deoxygenated blood within the RA, which subsequently flowed into both the LV and RV (Videos 5 and 6).

Based on the RA draining into both the RV and LV, and the anatomical location of the LAA relative to the obstruction of flow from the LA together with leftward deviation of the apical aspect of the IAS, a diagnosis of DORA was made.

Thoracic radiographs showed the presence of unstructured interstitial lung pattern, pulmonary venous congestion, and mild cardiomegaly (Figure 4).[1] These findings were supportive of cardiogenic pulmonary edema.

Serum biochemistry revealed the presence of mild azotemia (creatinine 136 μmol/L, ref. = 40-120; urea 14 mmol/L, ref. = 1.7-7.4). The

VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE, right parasternal long-axis, 4-chamber view, demonstrates leftward deviation of the IAS (*arrow*), which appears as a shelf on the dorsal aspect of the mitral valve. The RA communicates with both ventricles, and there is a small ostium primum septal defect (*) resulting in obstruction to the left atrial outflow.

Video 2: Two-dimensional TTE, left apical 4-chamber view, without (*left*) and with (*right*) color-flow Doppler demonstrates the flow obstruction to LA outflow caused by the leftward deviation of the IAS (*arrow*) and ostium primum defect. Interatrial communication (*) is also confirmed by the color-flow Doppler where aliasing flow across the left atrial outflow obstruction is discernible.

Video 3: Two-dimensional TTE, right parasternal short-axis view at the level of the mitral valve demonstrates the mitral valve leaflets are thickened and the valve has a trifoliate structure (*arrow*).

Video 4: Two-dimensional TTE, oblique short-axis view including the mitral valve with color-flow Doppler (aligned for assessment of mitral regurgitation) demonstrates moderate systolic mitral regurgitation into the RA with aliasing flow from the primum ASD during diastole. *IVS,* Interventricular septum; *LVFW,* left ventricular free wall; *MV,* mitral valve.

Video 5: Two-dimensional TTE, right parasternal long-axis 4-chamber view with color-flow Doppler demonstrates the eccentric jet of systolic mitral regurgitation entering the RA together with a continuous left-to-right shunt across the restrictive primum ASD. Diastolic laminar flow from the RA into the LV can also be seen. (Video playback is 50% actual speed to better appreciate the movement of blood flow).

Video 6: Two-dimensional TTE, left apical 4-chamber view with color-flow Doppler demonstrates the continuous high-velocity flow pattern across the ostium primum defect into the LV and an eccentric jet of mitral regurgitation into the RA. Normal diastolic flow from the RA appears to enter both the RV and the LV. (Video playback is 50% actual speed to better appreciate the movement of blood flow).

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

cat received intravenous furosemide boluses of 1 mg/kg as required until the respiratory rate stabilized at approximately 28 rpm with no effort; then the cat was transitioned onto oral diuretics (furosemide 2 mg/kg 3 times per day). Two days later blood pressure and azotemia were reported to be stable; therefore an angiotensin-converting enzyme inhibitor (benazepril 0.5 mg/kg once per day) was added to the treatment plan and the cat was discharged.

The cat was reported to be stable on heart failure treatment for 3 months, when sudden death occurred.

Postmortem examination confirmed a diagnosis of DORA (Figure 5). The dilated RA communicated directly through a broad outflow with both the RV and LV. The IAS was deviated to the left.

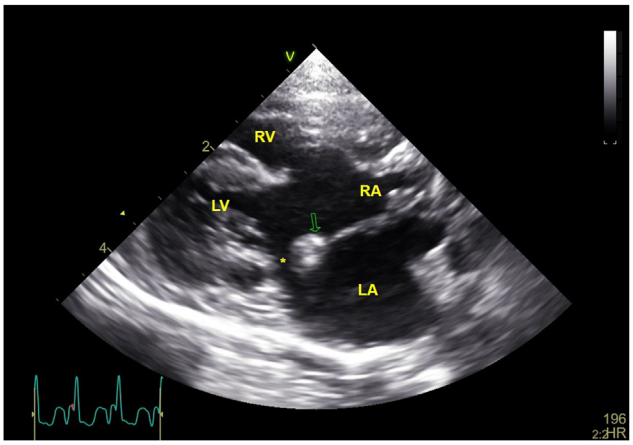

Figure 1 Two-dimensional TTE, right parasternal long-axis 4-chamber view, demonstrates leftward deviation of the IAS (*arrow*) that appears as a shelf on the dorsal aspect of the mitral valve. The RA communicates with both ventricles, and there is a small ostium primum septal defect (*) resulting in obstruction to the left atrial outflow.

The interventricular septum was hypertrophied but otherwise oriented normally. A narrow (~2 mm diameter) outflow from the LA to the ventral chamber (extension of the RA) was present. Two AV valves were present, as expected.

DISCUSSION

Atrioventricular septal defects, formerly called endocardial cushion or AV canal defects, result from maldevelopment of the AV septum. They can represent a range of complex congenital defects, including DORA, an abnormality that has seldom been reported in cats.[2-4] Double-outlet RA is a rare cardiac anomaly both in humans and animals. It describes the RA draining into both ventricles. Van Mierop[5] initially described this condition as a variant of "endocardial cushion defect" resulting from the extreme malalignment of the atrial septum. Indeed, the apical border of the septum primum fuses to the left lateral endocardial cushion near the left AV junction, causing LA outflow obstruction, instead of the inferior and superior cushions fusing over the interventricular septum.[6] This IAS deviation also results in separation of the supravalvular region into 2 compartments: a high-pressure LA (pressure proportional to the severity of the obstruction) and a low-pressure RA (that incorporates the entire RA and part of what anatomically should have been the remaining part of the LA).[2-5] Hence, in DORA cases the RA is the only atrium that drains directly into both the RV and LV. The RA receives blood from the cranial and caudal vena cava and from the LA through an ostium primum defect. This results in a mixture of oxygenated and nonoxygenated blood in the low-pressure compartment, which will then be directed to the pulmonary and systemic circulation.[2]

Specific features should be present to make a diagnosis of DORA[7] when this is due to abnormality of the IAS, as in this case. First, the lower portion of the IAS is deviated laterally and inserts into the left atrial free wall, extending as a shelf of tissue above the mitral valve, causing obstruction of the LA outflow and drainage of the RA directly into the LV. The second feature of DORA is the presence of a true LA chamber receiving all 4 pulmonary veins with a normally located

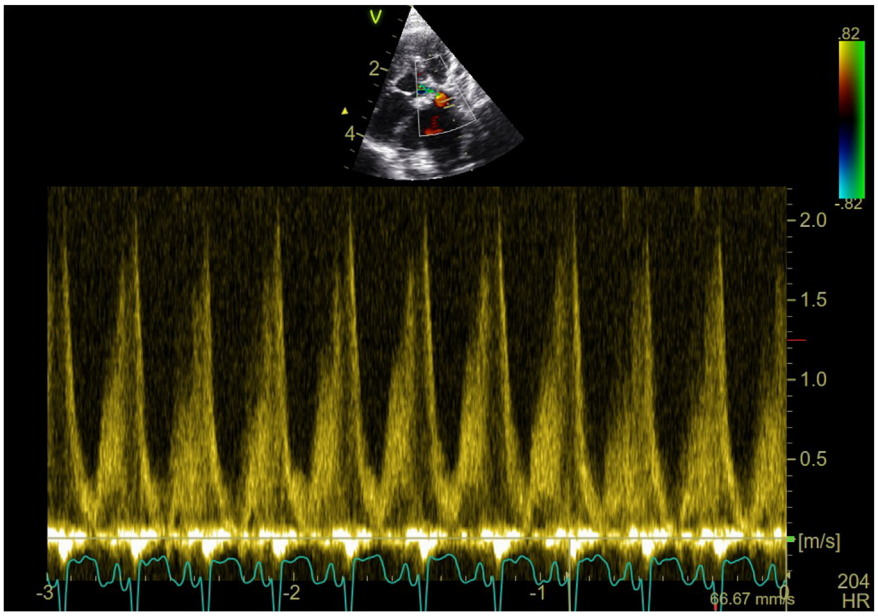

Figure 2 Two-dimensional TTE, left apical 4-chamber view (aligned for assessment of LA outflow), pulsed-wave spectral Doppler, demonstrates increased blood flow velocity (∼1.7 m/sec) due to the leftward deviation of the IAS and the small ostium primum septal defect.

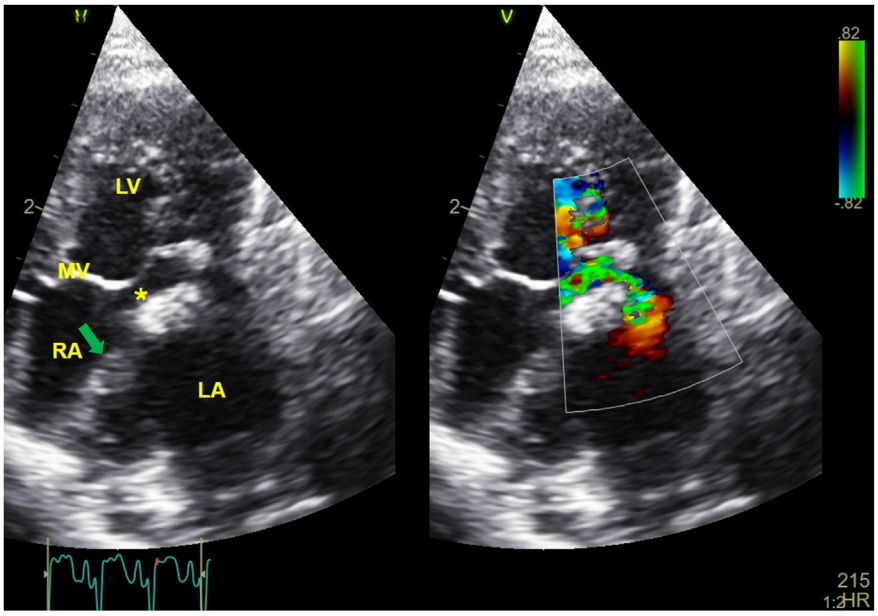

Figure 3 Two-dimensional TTE, left apical 4-chamber view without (*left*) and with (*right*) color-flow Doppler, demonstrates the flow obstruction to LA outflow caused by the leftward deviation of the IAS (*arrow*) and ostium primum defect (*). *MV*, Mitral valve.

LAA.[7] Double-outlet RA must be differentiated from cor triatriatum sinister and supravalvular mitral stenosis (SVMS).[7,8] A distinctive feature of cor triatriatum sinister is that the intra-atrial membrane is located above the orifice of the LAA; hence, the morphological LA does not communicate with the LAA, which consequently remains normal size.[8] This is not true in DORA.[2-8] Both DORA and SVMS are characterized by an enlarged LAA as the location of the intra-atrial shelf is more apical to the auricle orifice. In the case presented here, the absence of a normally oriented IAS and the presence of a partial AV septal defect supports a diagnosis of DORA rather than SVMS.[3] Trifoliate mitral valve is considered part of the spectrum of AV septal defects, and although this feature was not commented upon in the postmortem report, this was strongly suspected on echocardiographic assessment of the valve leaflets.[9,10]

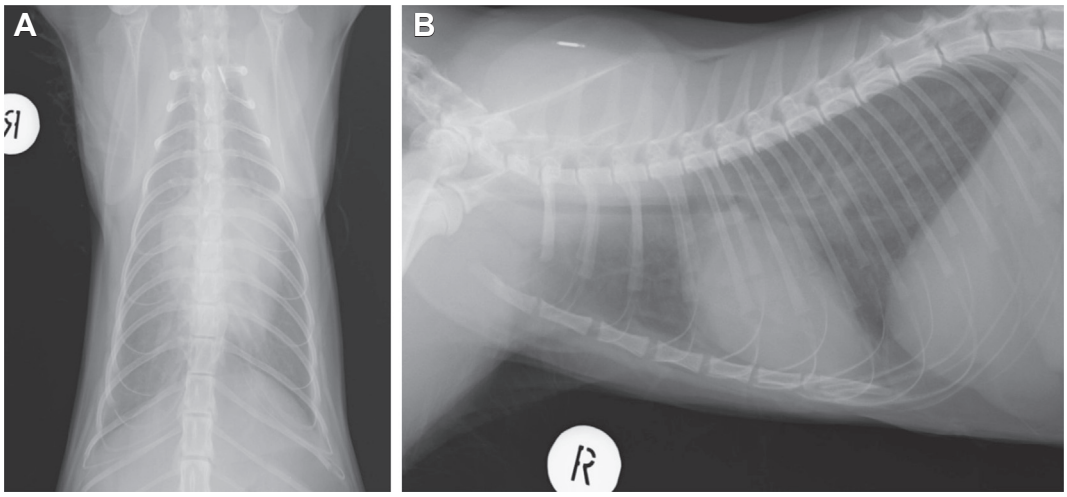

Figure 4 Dorsoventral (DV, **A**) and right lateral (RL, **B**) radiographic views of the thorax demonstrate a focal area of increased soft tissue opacity with an extrapleural sign between the cranial and caudal parts of the left cranial lung lobe (DV view) and a thin interlobar fissure between the cranial and caudal lung field (RL view). There is also a diffuse, mild interstitial lung pattern throughout the rest of the pulmonary parenchyma, and the pulmonary veins are dilated and taper peripherally. The cardiac silhouette is enlarged (vertebral heart size = 9 [reference interval, 6.7-8.1]).[1]

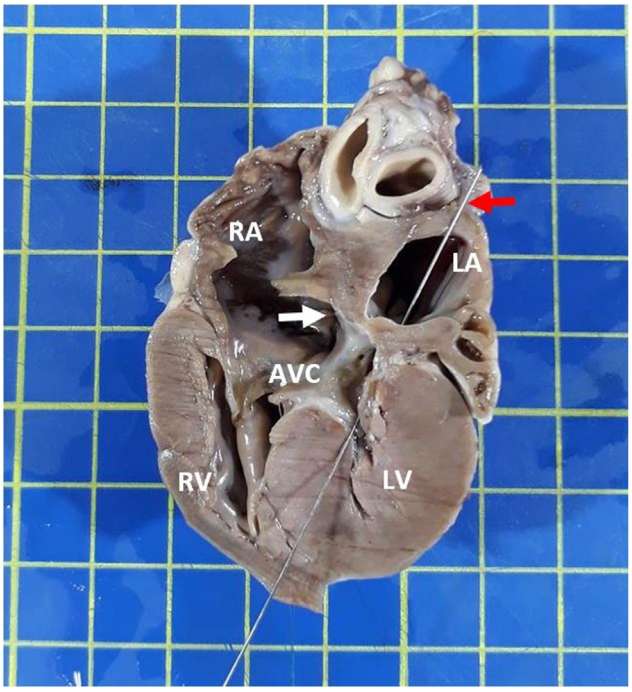

Figure 5 Gross pathology image of a sagittal plane of the heart with a metallic probe (*red arrow*) passing from the LA into the LV through a small ostium primum defect and part of the extended RA. The IAS (*white arrow*) deviates leftward and dorsal to the mitral valve. *AVC*, AV canal.

According to Perez-Martinez and colleagues,[11] DORA can be divided into 3 types based on the number of the AV valves present. Type A is described as having a single, straddling AV valve; type B is characterized as having components of 2 AV valves (which is most consistent with the case of this report); and type C is characterized as having 3 AV valves (2 tricuspid valves and 1 mitral valve). However, more recently the term *common-outlet RA* has been suggested to be more appropriate when describing the type A DORA reported by Perez-Martinez *et al.*[11,12] Furthermore, DORA is also classified on whether there is leftward atrial septal misalignment or rightward ventricular septal malalignment.[7-12]

Double-outlet RA has been reported in a total of 5 cats.[3,4] Clinical presentation of this condition in people is believed to be dependent on the size of the ASD and its associated anomalies. With a nonrestrictive ASD, the clinical findings are expected to be similar to those of a large ASD. With a restrictive ASD resulting in LA outflow obstruction, the clinical signs are often due to the development of left-sided congestive heart failure,[3,4,7] as in this case. Cyanosis or clinical signs associated with arterial desaturation are also possible and have been reported in patients affected by DORA.[5-7]

Based on the few reported cases in the veterinary literature, clinical presentation and age at diagnosis are variable. However, once signs of congestive heart failure occur, the prognosis is likely to be guarded.[3,4]

The overrepresentation of domestic shorthair cats affected by DORA likely reflects the high prevalence of this breed within the general population, instead of a true breed predisposition.[3,4]

One of the main hemodynamic consequences of the DORA defect is the right-to-left shunt (nonoxygenated blood from the RA shunting to the LV and systemic circulation).[5-7] The absence of cyanosis in the case reported here may be explained by the fact that the blood preferentially flowed from the RA to the RV rather than LV, in view of the high-pressure LV compared to the RV. Furthermore, the distinctive orientation of the IAS and location of the ASD facilitated flow of the oxygenated blood from the LA via the extension of the RA into the LV. The mitral regurgitant flow into the RA due to the levo-malalignment of the IAS resulted in mixing of oxygenated blood with nonoxygenated blood within the RA. This may have contributed to the absence of clinically significant

hypoxemia in this case. Hematology results (data not shown) were unremarkable in this cat, with a hematocrit at the higher end of the normal range (0.44 L/L; ref. = 0.26-0.44).

In human medicine, the treatment of choice for this cardiac defect is surgical correction, when feasible, and depends on the anatomical features of the ASD and the ventricles. In cases with restrictive ASD, intervention may be required to establish an AV connection and decrease the pressure within the anatomical LA and pulmonary veins. When the ASD is nonrestrictive, the clinical course is more benign, and the management depends on the severity of right-to-left shunting, AV valve structure, ventricular function, and associated anomalies. Surgical treatment involves complete excision and reconstruction of the abnormal IAS, with patch closure of the defect to separate the left and right AV valves.[7]

There are no studies in veterinary medicine focusing on possible surgical or interventional therapeutic options for cats affected by DORA. Instead, the treatment goal in those cats remains to control clinical signs of heart failure. However, with adequate and early diagnosis, a minimally invasive procedure for LA decompression (e.g., balloon septostomy at the level of the ASD orifice or transseptal puncture) or surgical reconstruction could be considered in some cases.

CONCLUSION

Double-outlet RA is a rare congenital cardiac defect that has been sporadically reported in people and cats. Although specific diagnostic criteria have been described for this defect, different pathoanatomical variations can make the diagnosis challenging. Clinical presentation and prognosis are variable and largely dependent on the anatomical and physiological characteristics of the defect, the size of the ASD, and the time of onset of clinical signs.

CONSENT STATEMENT

The authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

FUNDING STATEMENT

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with the ARRIVE guidelines and with the U.K. Animals (Scientific Procedures) Act, 1986 and associated guidelines, EU Directive 2010/63/EU for animal experiments, or the National Research Council's Guide for the Care and Use of Laboratory Animals.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2023.09.001.

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