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Short Communication

Bidirectional flow across a perforate cor triatriatum dexter in a dog with concurrent pulmonary, tricuspid, and mitral valve dysplasia☆,☆☆

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Abstract A 10-week-old male intact mixed breed dog presented for evaluation of suspected right-sided congestive heart failure. Echocardiographic imaging revealed a perforate cor triatriatum dexter (CTD), along with pulmonary valve stenosis and

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tricuspid and mitral valve dysplasia. In typical CTD cases, there is unidirectional blood flow across the dividing membrane, from the caudal into the cranial right atrial chambers. Owing to right-sided pressure alterations caused by the concurrent valvar defects, color Doppler imaging demonstrated bidirectional flow across the CTD membrane.

Abbreviations

CTD	cor triatriatum dexter
caRA	caudal right atrium
crRA	cranial right atrium
RA	right atrium

A 10-week-old male intact mixed breed dog, weighing 12.5 kg, presented to the Ohio State University Veterinary Medical Center for evaluation of suspected right-sided congestive heart failure. At presentation, physical examination abnormalities included the following: severe abdominal distention with a palpable fluid wave, bilateral jugular venous distension, mild tachypnea and dyspnea (respiratory rate of 80 breaths/minute), a grade V/VI right apical systolic heart murmur, and a grade IV/VI left basilar systolic heart murmur. Congenital heart disease was suspected, and a transthoracic M-mode, two-dimensional, and Doppler echocardiographic study were performed to characterize the heart disease.

Image interpretation

Images and video cine-loops were recorded from the left apical imaging window (Fig. 1, Video 1). Severe right atrial (RA) enlargement, moderate mixed eccentric and concentric right ventricular hypertrophy, a dilated coronary sinus, and multiple congenital heart defects were noted. A thin, perforate membrane was observed dividing the RA into a cranial RA (crRA) and caudal RA (caRA), consistent with cor triatriatum dexter (CTD). The dilated coronary sinus emptied into the caRA. The tricuspid valve was severely malformed characterized by thickened leaflets with restricted motion, diastolic doming, and incomplete coaptation during systole. The mitral valve was also malformed, with thickened leaflets and mild systolic prolapse. The left atrium and left ventricle were small and underfilled. Although not pictured, the pulmonary valve annulus was hypoplastic with infundibular narrowing and thickened immobile leaflets. Flow across the pulmonary valve was turbulent with a peak velocity of 3.6 m/s, suggesting a peak systolic transvalvular pressure gradient of 53 mmHg consistent with moderate pulmonary stenosis.

Doppler echocardiographic images from the left apical imaging window revealed valvar tricuspid stenosis with a mean diastolic pressure gradient of 7.5 mmHg, a peak early diastolic inflow velocity of 1.4 m/s, a prolonged pressure half-time of the E-F slope (168 ms [normal $<$ 50 ms [1]), and late diastolic inflow velocity of 2.1 m/s (Video 2). High-velocity tricuspid regurgitation was also apparent with peak velocity of 4.3 m/s, suggesting a right ventricular-to-RA pressure gradient of 74 mmHg. A small eccentric jet of mitral regurgitation was seen. Interrogation of the CTD membrane demonstrated bidirectional flow characterized as antegrade flow from the caRA into the crRA during systole and most of diastole (Fig. 2A). During late diastole (coincident with atrial contraction), CTD flow was reversed and traversed from the crRA into the caRA (Fig. 2B). Color M-mode imaging demonstrated the relative timing of the bidirectional CTD flow (Fig. 3).

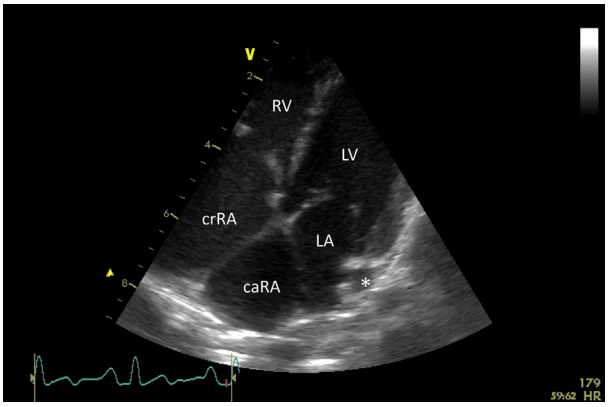

Fig. 1 Two-dimensional echocardiographic image from the left apical imaging window demonstrating severe enlargement of the right atrium. A thin, perforate membrane is seen separating the right atrium into a cranial right atrium (crRA) and caudal right atrium (caRA), consistent with the cor triatriatum dexter. This image highlights associated congenital defects including thickened malformed mitral and tricuspid valve leaflets between the left atrium (LA) and the left ventricle (LV) and crRA and right ventricle (RV), respectively. *Dilated coronary sinus.

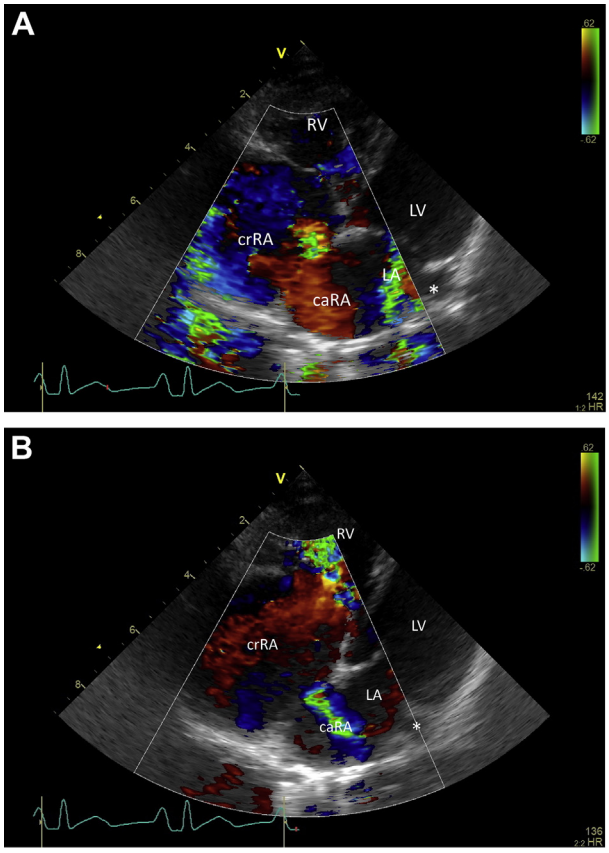

Fig. 2 Two-dimensional and color Doppler echocardiographic image from the left apical imaging window obtained during systole (A) and during late diastole after atrial contraction (B). In (A), antegrade blood flow is visualized from the caudal right atrial (caRA) into the cranial right atrial (crRA) chamber. Turbulent blood flow in the left atrium (LA) representing mitral regurgitation is also demonstrated. In (B), the flow across the membrane has reversed with flow from the crRA now entering the caRA. Turbulent inflow across the tricuspid valve as a result of tricuspid valve stenosis is also observed. RV, right ventricle; LV, left ventricle. *Dilated coronary sinus.

The puppy was euthanized, and a complete necropsy was performed confirming the presence of the congenital heart defects seen on the echocardiogram including the CTD and dysplasia of both atrioventricular and pulmonary valves, as demonstrate in Fig. 4A—D. Direct attachment of the papillary muscles to irregularly thickened tricuspid valve leaflets was observed. The diameter of the pulmonary outflow tract was narrowed and irregular, and the pulmonary valves were thickened and fused. The mitral valve leaflets were thickened with small 1 mm nodules along the free edge. A fossa ovalis was noted with no evidence of interatrial communication. The coronary sinus emptied into the caRA, and there was no evidence of a persistent left cranial vena cava. Two and half liters of ascitic fluid were removed from the peritoneal space. Marked chronic passive congestion was noted in the liver.

Discussion

Cor triatriatum dexter is a rare congenital cardiac anomaly defined by abnormal septation within the RA [2,3] and a fibrous or fibromuscular membrane dividing the RA into cranial and caudal chambers that communicate to a varying degree depending on the size of any perforations within the membrane [4]. In a typical CTD, because of the obstructive intra-atrial membrane, there is a higher pressure in the caRA than in the crRA throughout the cardiac cycle, typically resulting in continuous flow across the membrane from the caRA into the crRA [5]. In this dog, passive venous return resulted in blood flow from the caRA into the crRA during much of the cardiac cycle. During atrial systole, however, a transient increase in crRA pressure resulted in reversed flow across the membrane causing an atypical color and spectral Doppler appearance to this dog's CTD.

Although infrequent, CTD is well described in canine case reports [6—9]. The diagnosis is suspected from the two-dimensional imaging, and contrast echocardiography with agitated saline injected into the cranial and caudal peripheral vein has been suggested as a method to confirm this anatomy [10]. Contrast echocardiography was not pursued in this dog, but the reverse shunt flow across the CTD membrane may have altered interpretation of a contrast study as a cranial injection would have opacified the caRA and the crRA because of bidirectional flow between the chambers. Notably, lack of contrast echocardiography in this patient is a limitation of this case report.

Interventional management with balloon dilation or intravascular stenting of the obstructive membrane has been documented as potential therapeutic options in dogs, although efficacy depends on the presence or absence of concurrent congenital anomalies [6—9]. Balloon dilation of the CTD was offered in this dog, together with current tricuspid and pulmonary valvuloplasty, but was declined by the client.

In the dog, a CTD may be associated with additional congenital anomalies affecting the right heart including interatrial and interventricular communications, double-chambered right ventricle, ventricular hypoplasia, pulmonary valve stenosis, Ebstein's anomaly, and tricuspid valve

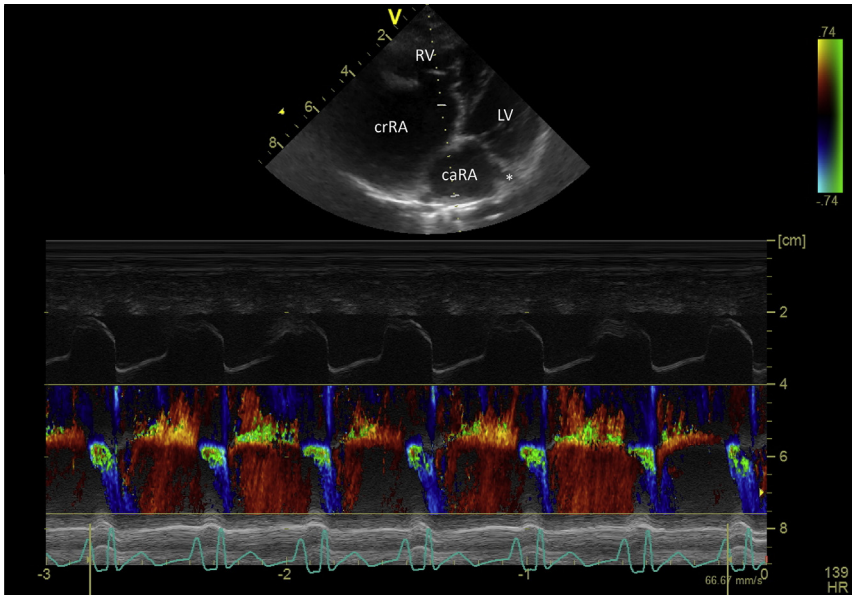

Fig. 3 Two-dimensional and color Doppler M-mode echocardiographic image from the left apical imaging window. Bidirectional flow across the cor triatriatum membrane is demonstrated here with reversed flow from the cranial right atrium (crRA) to the caudal right atrium (caRA) during atrial contraction (blue signal) and typical flow from caRA into the crRA throughout the remainder of the cardiac cycle (red signal). RV, right ventricle; LV, left ventricle. *Dilated coronary sinus.

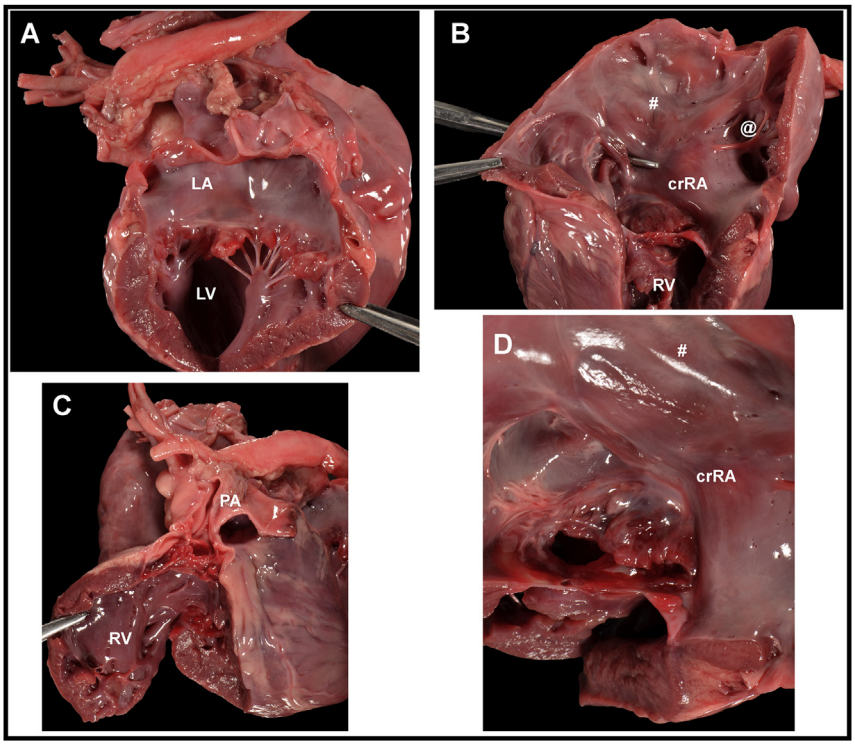

Fig. 4 Gross anatomical images from necropsy. (A) Left-sided view of the opened left atrium (LA) and left ventricle (LV) demonstrating mitral valve dysplasia characterized by thickened, nodular leaflets with short chordal attachments (B) right-sided view of the opened cranial right atrial (crRA) chamber and right ventricle (RV) demonstrating the probe-patent cor triatriatum dexter membrane, dilated crRA and right auricle (@), and an intact fossa ovalis (#) (C) right-cranial view of the opened RV outflow tract and main pulmonary artery (PA) demonstrating pulmonary valve stenosis with post-stenotic dilation of the main pulmonary artery (D) en face view of the dysplastic tricuspid valve demonstrating severely thickened and nodular leaflets with a stenotic orifice, dilated crRA chamber, and an intact fossa ovalis (#).

dysplasia [2—4,6,8,9]. In the dog of this report, multiple other anomalies were present including tricuspid and mitral valve dysplasia, and a hypoplastic and stenotic pulmonary valve. Retrospective studies evaluating congenital cardiac disease in dogs have found that multiple defects, as in the dog of this study, are present in roughly 14% of cases [11].

In this dog, severe valvar abnormalities led to elevated pressures within the right heart. These right-sided pressure alterations caused bidirectional blood flow across the CTD membrane, resulting in an atypical flow profile for this condition.

Conflict of interest statement

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

Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.jvc.2018.12.002.

Video table		
Video 1	Two-dimensional echocardiographic video of the left apical parasternal 4 chamber view.	Note the severely dilated right atrium and the concentric and eccentric hypertrophy within the right ventricle (RV). The cor triatriatum dexter membrane is seen dividing the right atrium into the cranial (crRA) and caudal (caRA) portions. The tricuspid and mitral valve leaflets are thickened with diastolic doming and inappropriate coaptation. Left ventricle (LV), left atrium (LA), dilated coronary sinus (*).
Video 2	Two-dimensional and color Doppler echocardiographic video of the left apical parasternal 4 chamber view.	Bidirectional flow across the cor triatriatum dexter membrane is demonstrated here. Also, note the tricuspid valve regurgitation and stenosis as well as mitral valve regurgitation. Cranial right atrium (crRA), caudal right atrium (caRA), right ventricle (RV), left ventricle (LV), left atrium LA), dilated coronary sinus (*).

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