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Cardiovascular Images

Cor triatriatum sinister in a dog[☆]

P. Castagna, DVM, G. Romito, DVM, PhD, M. Baron Toaldo, DVM, PhD[*]

Department of Veterinary Medical Sciences, Alma Mater Studiorum — University of Bologna, Italy

Abstract This report describes the transthoracic and transesophageal echocardio-graphic features of cor triatriatum sinister in an asymptomatic 6-year-old male French bulldog. Although cor triatriatum sinister represents a well-known and widely described cardiac malformation in humans, its description in the canine population is rare. In this clinical case, non-invasive echocardiographic techniques were helpful in visualizing and characterizing the lesion, allowing a valuable assessment of the malformation, and its hemodynamic consequences.
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Introduction

A 6-year-old, male intact, French bulldog weighting 14 kg was referred to the Veterinary Teaching Hospital of the University of Bologna for surgical resection of a low-grade cutaneous mast cell tumor. The dog appeared otherwise healthy. Specifically, cardiac auscultation revealed a regular rhythm with a heart rate of 136 beats/minute; no heart murmur could be detected. Femoral pulse quality was good,

and the examination of jugular veins showed no abnormalities. Thoracic radiographs were obtained as part of neoplastic disease staging.

Image interpretation

Thoracic radiography

This is a survey radiographic study of the thorax of a French bulldog, obtained in two perpendicular projections. On left lateral recumbency (Fig. 1A), a deformation of the profile of the left atrium (LA) is evident (white dotted line). No other alterations of the cardiac silhouette were noticeable in the ventrodorsal projection (Fig. 1B). Given the radiographic abnormality of the LA, preanesthetic echocardiography was recommended. Complete transthoracic echocardiography (TTE), including two-dimensional, M-mode, and Doppler analysis, was performed according to a standard technique [1] using an ultrasound unit[a] equipped with multifrequency phased array transducers and with continuous ECG monitoring.

Two-dimensional TTE

On the right parasternal long-axis view (Fig. 2A, Video 1), the atrioventricular valves, as well as dimensions and systolic function of the left ventricle (LV), were morphologically normal. Within the LA, an interrupted linear hyperechoic structure compatible with a fenestrated membrane was identified. This anomalous membrane appeared to transect the LA, partitioning it into two discrete chambers. The proximal portion (the accessory atrial chamber [AC]) appeared to be located dorsocaudally, whereas the distal portion (the true LA) was in direct communication with the LV, through the mitral valve orifice, and contained the atrial septum. Some pulmonary veins were draining into the AC (star), whereas the others were connected to the true LA (asterisks). The region of

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[a] iE33 ultrasound system, Philips Healthcare, Monza, Italy.
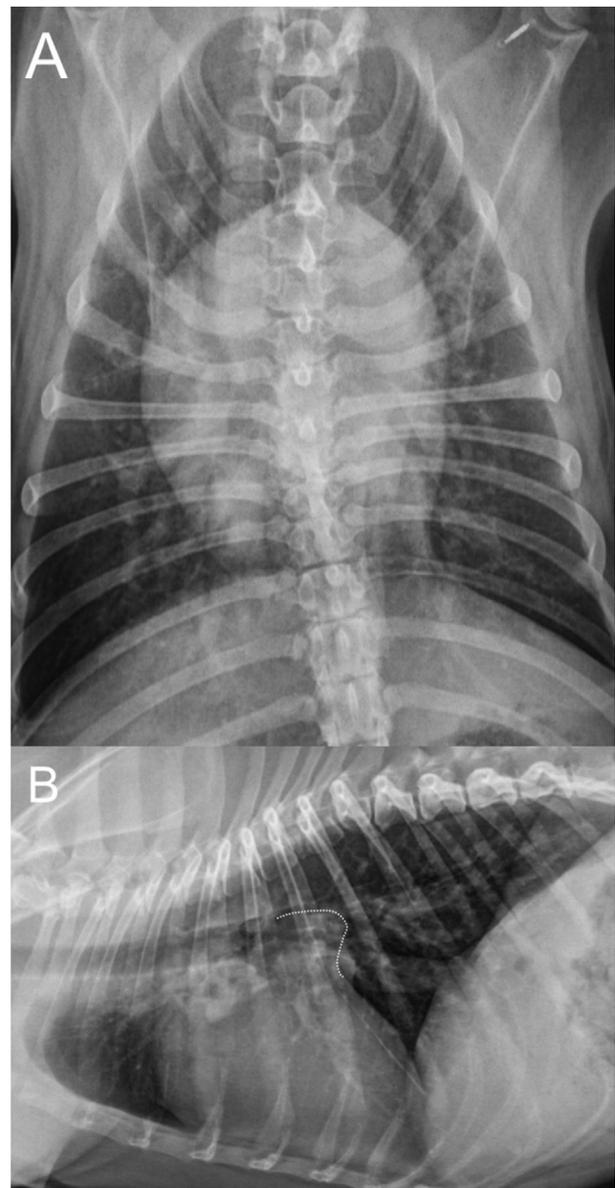
Fig. 1 (A) Left lateral and (B) ventrodorsal thoracic radiographs from a French bulldog. (A) Note the evidence of left atrial dilation on lateral recumbency. The radiographic border of the caudodorsal aspect of the left atrium is outlined (white dotted line) for a better assessment of such a radiographic change. No additional radiographic abnormalities are evident.

the fossa ovalis appeared particularly thin but intact based on color Doppler interrogation on TTE. The short-axis view at the level of the cardiac base (Fig. 2B) provided a better visualization of the AC and its direct connection to some pulmonary veins (white stars). The left auricle was, instead, part of the true LA. Combining information obtained from both echocardiographic views, the right middle lobe and the right cranial lobe pulmonary veins were judged to drain into the LA,

Fig. 2 Two-dimensional transthoracic echocardiographic image obtained from a right parasternal long-axis view. (A) Note the thi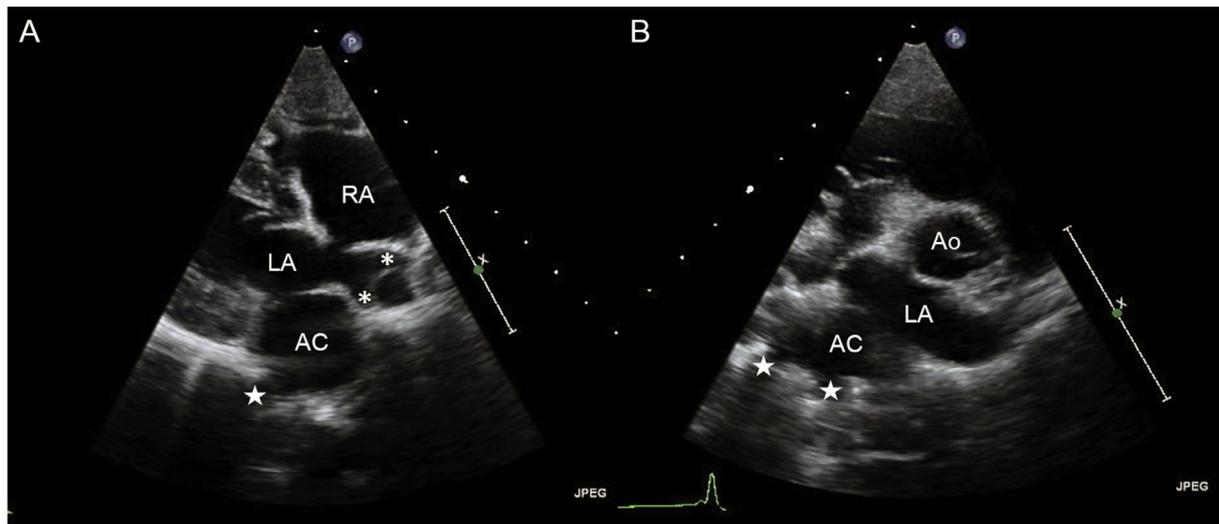n perforated membrane partitioning the left atrium (LA) into two chambers: an accessory chamber and the true LA, which includes the mitral valve annulus and the interatrial septum. (B) Right parasternal short-axis view at the level of the cardiac base. Note that the accessory chamber lies proximally to the left atrial appendage that is in direct continuation with the true LA. In both views, some pulmonary veins drain into the true LA (white asterisks), whereas other veins enter the accessory chamber (white stars). AC: accessory atrial chamber; Ao: aorta; LA: left atrium; LV: left ventricle; RA: right atrium.

whereas the AC appeared to receive the blood from the left caudal and left cranial lung lobes, the accessory lobe, and the right caudal lobe pulmonary veins [2]. Computed tomography was not performed to further characterize the pulmonary venous drainage.

Color Doppler TTE

On color Doppler analysis obtained through a left apical four-chamber view, a continuous forward blood flow across the membrane's orifice was evident. The true LA was receiving blood coming from both the AC, with a turbulent jet directed toward the anterior mitral valve leaflet, and the pulmonary veins likely draining the right cranial and middle lung lobes. The remainder of the echocardiographic study was normal. In light of the aforementioned findings, cor triatriatum sinister (CTS) was diagnosed.

One week after TTE, the surgical resection of the cutaneous mast cell tumor was performed without any complications. Immediately after surgery, still under general anesthesia, transesophageal echocardiography (TEE) was carried out using a dedicated probe,[b] to better characterize the cardiac malformation (Video 2).

[b] X7-2t transesophageal phased array transducer, Philips Healthcare, Monza, Italy.

Transesophageal echocardiography: two-dimensional and color Doppler analysis

Transesophageal echocardiography was performed with the dog positioned in right lateral recumbency following standard techniques [3]. Figure 3A shows a middle-position TEE view, with the cursor angle set at 141°, obtaining a modified four-chamber view. The AC is visible on the upper right part of the image with the perforated membrane identified by the star. The interatrial septum was normal in appearance. A slight rotation of the cursor to 116°, allowed visualization of the LV outflow tract and aorta (Fig. 3B, Video 3). Color Doppler analysis clearly showed a continuous turbulent blood flow across the membrane's orifice directed straight toward the anterior mitral valve leaflet. In addition, mild systolic mitral valve regurgitation was evident and eccentrically directed.

Transesophageal echocardiography: spectral Doppler and color M-mode analysis

The continuous-wave Doppler analysis of the transmembrane blood flow revealed a continuous systodiastolic flow with a maximal velocity during ventricular diastole (velocity, 1.8 m/s; pressure gradient, 13 mmHg) and a minimal velocity during ventricular systole (velocity, 1.3 m/s; pressure
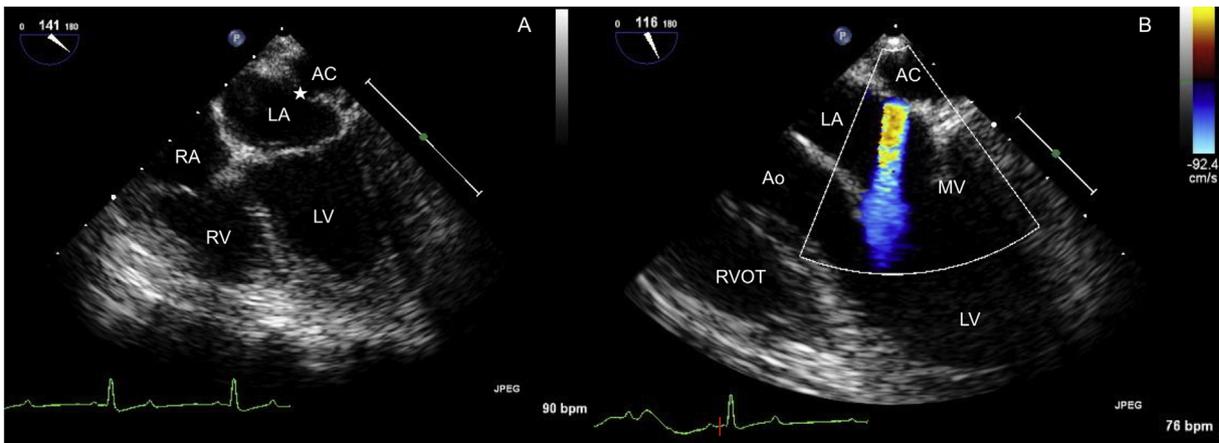
Fig. 3 (A) Transesophageal echocardiographic view obtained with the probe placed in the middle position and showing a modified four-chamber view. Note the fenestrated membrane (white star) dividing the left atrium from the accessory chamber. (B) Color Doppler analysis of the malformation from a left ventricular outflow tract view. Note the continuous turbulent blood flow across the membrane's orifice with a restrictive behavior and directed toward the anterior mitral valve leaflet. The numbers on top left of the images (141° and 116°) represent the degree of rotation of the crystal of the transesophageal probe. AC: accessory atrial chamber; Ao: aorta; LA: left atrium; LV: left ventricle; MV: mitral valve; RA: right atrium; RV: right ventricle; RVOT: right ventricular outflow tract.

gradient, 7 mmHg). A short notch on the spectral profile occurred in concomitance with mitral valve closure (Fig. 4A). The maximal pressure into the AC was estimated to be around 20 mmHg, assuming a LA systolic pressure of 7—8 mmHg.

The color Doppler and the M-mode modalities were combined to display mechanical events in addition to flow events simultaneously in the AC, LA, and LV (Fig. 4B). The cursor line was superimposed to the turbulent jet; therefore, on the M-mode trace, the LA tier was occupied by an aliased/mainly yellow-colored flow. The movement of the anterior mitral valve leaflet set the passage of blood into the LV that filled during the early and late diastolic phase (blue bands in the bottom tier). The red jet represented a mild early systolic mitral valve regurgitant jet (arrowhead). The narrow blue band between the AC and LA tiers reflected the small area of isovelocity of the turbulent jet on the AC side of the perforated membrane.

After completion of the TEE, the dog recovered uneventfully from anesthesia and was discharged
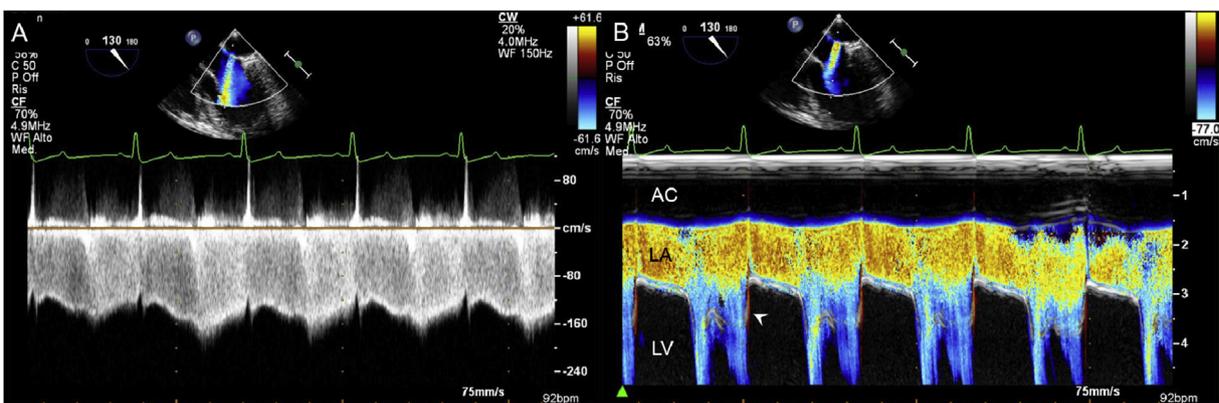
Fig. 4 (A) Continuous-wave Doppler analysis of the transmembrane blood flow obtained from the same image as Fig. 3B. Notice the continuous blood flow across the perforated membrane that is directed distally toward the left atrium. The minimal velocity is recorded during ventricular systole, whereas the maximal flow velocity across the membrane occurs during ventricular diastole, when the mitral valve is open. (B) Color M-mode analysis shows the distribution of the blood flow across the different left-sided chambers and their timing. The turbulent jet across the membrane invades the left atrium during the entire cardiac cycle, whereas the LV is filled normally during diastole. Mild mitral regurgitation is visualized in early systole as a thin red jet (arrowhead). AC: accessory atrial chamber; LA: left atrium; LV: left ventricle.

on the same day. No therapies were recommended for the cardiac abnormality, but echocardiographic rechecks were suggested to monitor the malformation over time.

Discussion

Cor triatriatum sinister is a rare cardiac malformation in which the LA is divided into two compartments by an anomalous membrane. Consequently, three atrial chambers can be identified: the right atrium, the actual true LA, and the AC connected to the LA (hence, the term 'triatriatum') [5]. The membrane that transects the LA varies significantly in size and shape; it may appear similar to a diaphragm or be funnel shaped, entirely intact (imperforate), or contain one or more fenestrations with different diameters [5]. In humans, CTS represents only 0.1—0.4% of all congenital cardiac malformations and may be associated with other cardiac defects in as many as 50% of cases, especially atrial septal defects and abnormalities of the pulmonary venous return [5]. As opposed to the more frequently described cor triatriatum dexter in dogs and CTS in cats, CTS is rare in the canine species. Thus far, only two cases have been published, both presented as a solitary malformation associated with signs of congestive heart failure [6,7]. Specifically, in one case (a 3-year-old male French bulldog), the diagnosis was made exclusively by TTE when clinical signs of lung edema developed [6], whereas in the other one (a 5-year-old female poodle), evidence of CTS was obtained exclusively by postmortem examination [7]. Although several theories have been hypothesized in human medicine, the embryologic basis of CTS is still a controversial subject. To date, three main theories have been proposed: the 'malincorporation' (incomplete incorporation of the common pulmonary vein into the LA), the 'malseptation' (the septum subdividing the LA is the result of an abnormal overgrowth of the septum primum), and the 'entrapment' (the left horn of the embryonic sinus venosus entraps the common pulmonary vein, thereby, preventing its incorporation into the LA) [5]. In humans, several classifications of CTS have been suggested based on the communication between the AC and the other cardiac chambers, the quantity and morphology of fenestrations of the anomalous membrane, and concurrent anomalies of the venous return [8]. According to the classification proposed by Krabill and Lucas [9], the dog of this report had CTS with a C1A morphology, characterized by subtotal CTS where the AC connects to the LA and receives part of the pulmonary veins, whereas the remaining

pulmonary veins drain normally into the true LA. Human and canine patients with CTS may be asymptomatic, as in the case presented here, or show signs of left-sided congestive heart failure owing to the obstruction to the flow between the pulmonary venous system and the LV [5—7]. The development of clinical signs and the time of their occurrence mainly depend on the size and number of the fenestrations in the membrane as well as the presence of concurrent heart abnormalities [5]. In human medicine, several imaging techniques have been used to establish the CTS diagnosis, including two-dimensional and three-dimensional TTE and TEE, cardiac catheterization and selective angiography, computed tomography, and magnetic resonance imaging [8]. Traditionally, either in human and veterinary medicine, TTE has represented the first-line diagnostic modality because it is relatively inexpensive, widely available, non-invasive, and easy to perform. In addition, this imaging technique can identify the possible hemodynamic compromise associated with CTS, and it might reveal concomitant cardiac abnormalities [6—8]. In cases of CTS associated with multiple cardiac defects or patients with a peculiar chest conformation (e.g., obese human patients, brachycephalic canine breeds), TTE alone could lead to an inadequate visualization of the AC and of the abnormal membrane. In such cases, TEE has been demonstrated to represent a valuable complementary tool in people [8]. As compared with TTE, TEE offers superior visualization of cardiac structures because of the close proximity of the esophagus to the heart and lack of superimposition of the lungs, muscles, and bones. In addition, this proximity permits use of high-frequency imaging transducers that afford superior spatial resolution [3]. In the case described here, TEE allowed a better visualization of the LA anatomy, of the interatrial septum, and the abnormal membrane, with confirmation of the malformation and of the origin of the pathologic flow.

In symptomatic human patients, medical therapy can be initially set up to control the clinical signs of congestive heart failure, although the treatment of choice remains the surgical resection of the intra-atrial membrane [5]. Alternatively, minimally invasive percutaneous catheter balloon dilatation of the membrane can be considered [10]. To date, only the medical approach (e.g., furosemide and benazepril) has been reported in canine CTS [6,7], although either surgical resection and balloon dilation have been demonstrated to be effective in dogs with cor triatriatum dexter [11] and in cats with CTS [12]. Among the two therapeutic options, balloon dilation represents a less expensive and

dangerous approach for this malformation in small animals [13]. A hybrid technique, performed by inserting an inflatable balloon through the defect after thoracotomy approach to the LA, has also been successfully performed in one cat with CTS [14]. In asymptomatic human patients, no intervention is usually advised, but regular rechecks are planned over time to identify any progressive narrowing of the membrane's orifice, signs of venous congestion, or occurrence of pulmonary hypertension. In the dog from the present report, the elected approach was to monitor, given that the patient was asymptomatic and there was no evidence of pulmonary venous congestion. On the other side, the measured gradient across the membrane was about 20 mmHg. It might be speculated that this chronic increase in pressure might lead to a remodeling of the affected vasculature and possibly induce a chronic and clinically relevant pulmonary hypertension. Considering the overall case, a 'wait-and-see' approach was considered more appropriate, and an interventional treatment was offered as a plausible option in case the malformation became hemodynamically relevant.

In conclusion, this is the first report of a canine CTS extensively described with the use of TTE and TEE. The addition of another imaging modality such as TEE may help to further expand the characterization of this rare congenital defect.

Conflicts of Interest Statement

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

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Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jvc.2019.07.003.

Video table		
Video	Title	Description
1	Transthoracic echocardiographic video clip obtained from a right parasternal long-axis view in a French bulldog.	The left atrium is partitioned into two chambers, a proximal one (accessory chamber) and a true left atrium, that are divided by a perforated thin membrane. The pulmonary veins appear to drain partly into the accessory chamber and partly into the true left atrium. The other cardiac structures are normal.
2	Color Doppler transthoracic echocardiographic video clip obtained from a left apical four-chamber view.	Note the hyperechoic band traversing the LA consistent with the anomalous membrane of cor triatriatum sinister (left panel). On color Doppler imaging (right panel), there is continuous flow arising from the membrane's orifice and directed toward the anterior mitral valve leaflet. Intermittent blood flow arising from some pulmonary veins that directly communicate with the true left atrial cavity is observed.
3	Color Doppler transesophageal echocardiographic video clip obtained from a left apical four-chamber view with visualization of the left ventricular outflow tract.	A continuous, turbulent blood flow arising from the accessory atrial chamber and invading the true left atrium through the membrane orifice is evident. This flow is directed toward the anterior mitral valve leaflet. Note also the mild, eccentric, systolic blood flow across the mitral valve, consistent with mitral regurgitation.

AC: accessory atrial chamber; LA: left atrium.

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