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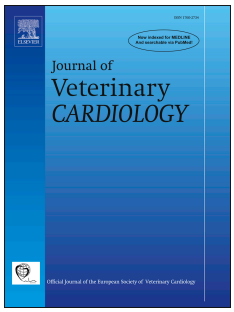

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Cor triatriatum sinister in a dog

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Short title: Cor triatriatum sinister

Abstract

This report describes the transthoracic and transesophageal echocardiographic features of a *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.

Abbreviation Table

AC	accessory atrial chamber
CTS	cor triatriatum sinister
LA	left atrium
LV	left ventricle
TEE	transesophageal echocardiography
TTE	transthoracic echocardiography

Keywords

Transesophageal echocardiography; cardiac malformation; left atrium; canine.

Introduction

A 6-year-old, male intact, French bulldog weighing 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:

Figure 1. Thoracic radiography

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 left atrial (LA) profile in evident (white dotted line). No other alterations of the cardiac silhouette were noticeable in ventrodorsal projection (Fig. 1B). Given the radiographic LA abnormality, pre-anesthetic 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 multi-frequency phased array transducers and with continuous ECG monitoring.

Figure 2 and Video 1. Two-dimensional transthoracic echocardiography

On right parasternal long axis view (Fig. 2A, Video 1), the atrioventricular valves were morphologically normal as well as the left ventricular (LV) dimensions and systolic function. 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 located dorso-caudally, while 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), while others were connected to the true LA (asterisks). The region of 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, while 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.

Video 2. Color Doppler transthoracic echocardiography

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 above-mentioned findings, a *cor triatriatum sinister* (CTS) was diagnosed.

One week after the 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.

Figure 3, Video 3. 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-chambers 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 visu alization 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. Additionally, a mild systolic mitral valve regurgitation was evident and eccentrically directed.

Figure 4. Transesophageal echocardiography – Spectral Doppler and color M-Mode analysis

The continuous-wave Doppler analysis of the transmembrane blood flow revealed a continuous systo-diastolic 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 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 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 the 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 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 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], while in the other one (a 5-year-old female Poodle) evidence of CTS was obtained exclusively by post-mortem 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 left atrium is the result of an abnormal overgrowth of *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, 1995 [9], the dog of this report had a CTS with a C1A morphology, characterized by a subtotal CTS where the AC connects to the LA and receives part of the pulmonary veins, while 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 due to the obstruction to the flow between the pulmonary venous system and the left ventricle [5-7]. The development of clinical signs and the time of their occurrence mainly depends 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-dimentional as well as 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. Additionally, this imaging technique can identify the possible hemodynamic compromise associated with the CTS and it might reveal concomitant cardiac abnormalities [6-8]. In cases of CTS associated with multiple cardiac defects or for 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. Additionally, 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 per-catheter balloon dilatation of the membrane can be considered [10]. To date, only the medical approach (e.g., furosemide, 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 thoracotomic 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.

Conflict of interest

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

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Footnotes

[a] iE33 ultrasound system, Philips Healthcare, Monza, Italy.

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

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Figure legends

Fig. 1: Left lateral (A) and ventro-dorsal (B) thoracic radiographs from a French Bulldog. Note the evidence of left atrial on lateral recumbency (A). The radiographic border of the caudo-dorsal aspect of the left atrium is outlined (white dotted line) for a better assessment of such radiographic change. No additional radiographic abnormalities are evident.

Fig. 2: Two-dimensional transthoracic echocardiographic image obtained from a right parasternal long axis view. Note the thin 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 (A). Right parasternal short axis view at the level of the cardiac base (B). Note that the accessory chamber lays 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) while other veins enter the accessory chamber (white stars). AC: accessory atrial chamber; Ao: aorta; LA: left atrium; LV: left ventricle; RA: right atrium.

Fig. 3: A. Transesophageal echocardiographic view obtained with the probe placed in middle position and showing a modified four-chamber view (A). Note the fenestrated membrane (white star) dividing the left atrium from the accessory chamber. Color Doppler analysis of the malformation from a left ventricular outflow tract view (B). 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.

Fig. 4: A. Continuous-wave Doppler analysis of the transmembrane blood flow obtained from the same image as Figure 3B (A). 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, while the maximal flow velocity across the membrane occurs during ventricular diastole, when the mitral valve is open. Color M-Mode analysis shows the distribution of the blood flow across the different left-sided chambers and their timing (B). The turbulent jet across the membrane invades the left atrium during the entire cardiac cycle, while 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.

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 in two chambers, a proximal one (accessory chamber) and a true left atrium, which 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. AC: accessory atrial chamber; LA: left atrium.
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	A continuous, turbulent blood flow

	echocardiographic video clip obtained from a left apical four-chamber view with visualization of the left ventricular outflow tract.	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.

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