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Letter to the Editor

Eisenmenger's Syndrome in a Cat with Ventricular Septal Defect (Ventriküler Septal Defektli Bir Kedide Eisenmenger Sendromu)

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Dear Editor,

Ventricular septal defect (VSD), a congenital cardiac malformation, is characterized by an abnormal communication between two ventricular chambers. Congenital heart defects (VSD, ASD and PDA) cause left-to-right (LR) cardiac shunt, and then right-to-left (RL) shunt when right ventricular pressure exceeds the left ventricular (LV) pressure, resulting in pulmonary artery hypertension (PH). That cardiac shunt switches to a cyanotic RL shunt from LR shunt due to PH, which is defined as Eisenmenger's syndrome, has a high risk factor for mortality [1]. Endothelin (ET) plays a key role of pulmonary vasoconstriction in the pathophysiology of Eisenmenger's syndrome. There is limited information on the diagnostic steps of Eisenmenger's syndrome in cats [1,2], and also no available data on the use of ET receptor antagonists in the treatment of this syndrome in dogs and cats. Thus, this case report aims to share practical knowledge on size detection of the VSD by two non-invasive techniques; echocardiography and three-dimensional computed tomographic angiography (3D-CTA) and treatment possibilities in non-operable feline patients with Eisenmenger's syndrome.

A British shorthair cat (1.5 year, female, and 2.5 kg) was referred from a small animal clinic to Animal Hospital with a history of lethargy, exercise intolerance, and respiratory distress during exercise for 3 months. Before admission, the cat was treated with furosemide for 5 days due to pulmonary oedema. In physical examination, body temperature and heart and respiratory rates were within reference ranges. Cyanotic mucous membranes were observed. Systolic cardiac murmur - grade 4/6 was auscultated over left and right 2-4 intercostal spaces. Thoracic radiography revealed enlarged heart size silhouette and pulmonary artery bulging with an alveolar pattern lung tissue appearance. ECG examination showed rS complexes and right axis deviation suggestive for right-sided cardiac enlargement. Complete blood cell count was non-specific (Hct: 43.4%, reference: 24-45%). Serum biochemistry profile showed pre-renal azotemia (BUN: 36 mg/dL, reference: 10-30 mg/dL; Cr: 1.3 mg/dL, reference: 0.3-1.8 mg/dL) and hyponatremia (137 mmol/L, reference: 142-164 mmol/L) due to diuretic administration.

Standard echocardiographic images showed severe right (2.22 cm) and left atrial enlargement (1.79 cm, reference 8-13 cm) with an increased left atrial-to-aortic diameter ratio (2.4, reference <1.5) on the right parasternal long and short axis views. M-mode measurements of the LV at diastole and systole showed that all geometric and functional parameters were within reference ranges; LV dimensions - 1.41 cm (1.2-1.8 cm) and - 0.50 cm (0.5-1.0 cm), interventricular septal thickness - 0.41 cm (0.3-0.5 cm) and - 0.5 cm (0.5-0.9 cm), LV post wall thickness - 0.46 cm (0.3-0.5 cm) and - 1.0 cm (0.4-0.9 cm) and fractional shortening - 30% (30-55%), respectively. Pulmonary and aortic Doppler flow velocities were 1.9 m/s (≤1.2 m/s) and 0.88 m/s (≤1.2 m/s), respectively. A 2.6 mm VSD was identified with RL shunting on color Doppler, with a maximum velocity of 2.3 m/s (estimated peak pressure gradient of 21.6 mmHg). Normally, since the pulmonary blood flow (Qp) is equal to the systemic blood flow (Qs), their ratio (Qp:Qs) is equal to one. Qp:Qs shows the direction and magnitude of the shunting occurring LR (greater than one) or RL (less than one) [3]. In this case, echocardiographic estimation of Qp:Qs

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(0.8) in the first day of diagnostic work-up was consistent with suspected shunt way. An agitated saline micro-bubble contrast study performed via the cephalic vein confirmed a RL shunt across the interventricular septum. Inlet VSD (atrioventricular septal defect) was confirmed by use of 3D-CTA *(Fig. 1)*, as reported in a previous case [4]. Based on these observations, the cat was diagnosed with VSD and suspected pulmonary vascular obstructive disease resulting in PH (Eisenmenger's syndrome). In this case, lack of polycythemia maybe due to relatively recent shunt reversal, in agreement with the acute decompensation before presentation, as reported earlier [2].

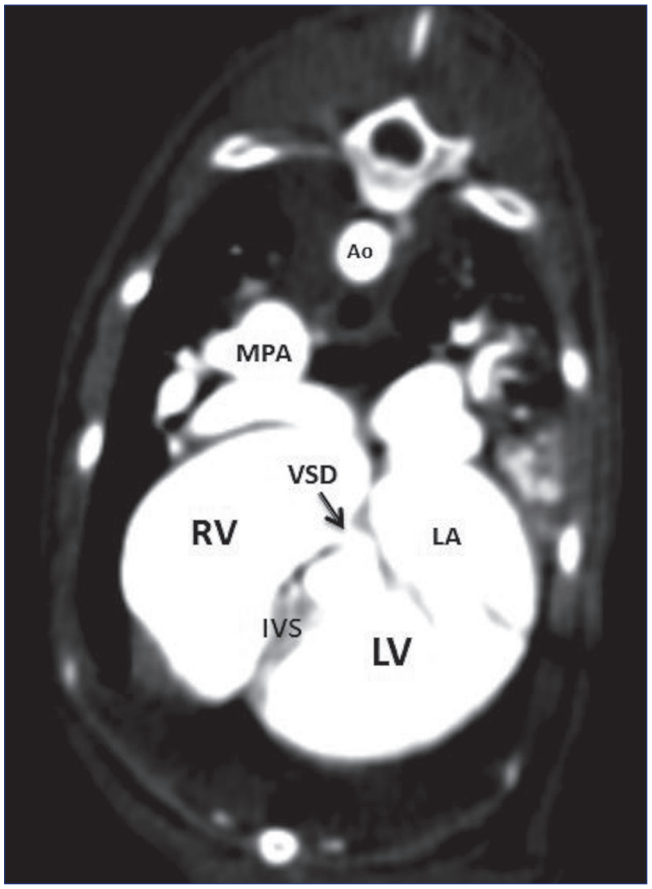

Fig 1. Three-dimensional computed tomographic angiography shows the presence of ventricular septal defect (VSD) just behind the atrioventricular valves between two ventricular chambers

Ao: aorta, RV: right ventricle, LA: left atrium, LV: left ventricle, IVS: interventricular septum, MPA: mean pulmonary artery

The cat was treated with an ACE-i drug (enelapril, 0.5 mg/kg 1x1, PO), diuretic (furosemide, 2 x 2 mg/kg, PO) and dietary salt restriction, to alleviate PH and reduce volume overload. Because exercise intolerance was stabile for two weeks despite to the treatment regimen, medical strategy was modified. Sildenafil (Revatio® 20 mg/tablet, Pfizer, 0.5 mg/kg, PO, q12 hr) was administered as an oral phosphodiesterase type V inhibitor acting preferentially to vasodilate arteries in the lung for two weeks, as suggested [1]. Since Eisenmenger's syndrome is associated with increased ET expression, patients may benefit from ET receptor antagonism. Thus, bosentan as an oral ET-1 receptor antagonist (Tracleer 125 mg/tablet, 3 mg/kg, PO, q12 h) was suggested with concomitant use of sildenafil citrate [5]. Although bosentan could not effective to produce dramatic changes in Doppler spectral pattern of flow through pulmonary artery and septal defect, it was well tolerated and improved the exercise capacity. Thus the cat was alert and not shown the clinical signs on the control examinations with two week intervals for 3 months.

In conclusion, high technological imaging systems such as 3D-CTA may help to confirm the diagnosis of congenital cardiac defects. VSD should be considered as a risk factor leading to PH (Eisenmenger's syndrome) in cats as reported in human medicine. Thus, in addition to traditional therapy (ACE-I and diuretic), sildenafil and/or bosentan may be used to alleviate clinical signs in these cases.

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