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Tetralogy of Fallot in an Alaskan malamute: a case report

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Abstract: A 3-month-old female Alaskan malamute was presented with a grade IV/VI systolic heart murmur, exercise intolerance, and anorexia. On examination, the dog was found to have cyanosis and inadequate growth. Echocardiography revealed a dextroposition of the aorta, a large ventricular septal defect, severe pulmonic stenosis (maximal velocity 4.77 m/s), and right ventricular hypertrophy. The dog was diagnosed with tetralogy of Fallot (TOF) and managed medically with atenolol 0.3 mg/kg SID. The incidence of TOF in dogs has been reported to be approximately 0.6 - 1%. This report is the first to encompass a complete blood count, thoracic radiographs, and echocardiography findings in an Alaskan malamute diagnosed with TOF in Thailand.

Keywords: tetralogy of Fallot, cyanosis, heart murmur, Alaskan malamute, congenital heart disease

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Introduction

Tetralogy of Fallot (TOF) is a rare congenital heart defect characterized by four congenital malformations comprised of ventricular septal defect (VSD), displacement of the aortic root on the right side, over the interventricular septum, a so-called overriding aorta (dextroposition of aorta), pulmonic stenosis (PS), and right ventricular (RV) hypertrophy. TOF is the most common cyanotic heart defect diagnosed in humans and animals (Apitz *et al*., 2009). This malformation has been described in animals such as dogs, cats, horses, sheep, pigs, cattle (Michaelson and Ho, 2000), and bears (Ågren *et al*., 2005). The prevalence of TOF in dogs has been reported approximately 2.3% of all canine congenital cardiac anomalies (Brambilla *et al*., 2020) and genetically transmitted in some dog breeds such as the Keeshond and English bulldog (Bonagura and Lehmkuhl, 1999). The condition is considered even rarer in the cat. In a few affected animals, TOF may be tolerated for years before clinical signs are noted. However, most affected dogs have severe exercise intolerance, cyanosis and sudden death is a common occurrence in this subset of patients (Beijerink *et al*., 2017). Survival time of dogs with untreated TOF is not very high and the median age at the time of (cardiac-related) death has been reported to be approximately 23.4 months (Chetboul *et al*., 2016). Dogs with the successful placement of the modified Blalock-Taussig shunt (mBT shunt procedure) quality of life appeared improved and survived long term can live up to 6 years (Brockman *et al*., 2007).

Materials and methods

A 3-month-old, female Alaskan malamute was brought to the animal hospital with a prominent heart murmur, exercise intolerance. The patient was thin and smaller than her littermates but was otherwise well developed with a body condition score of three out of nine (body weight 8.9 kg). The patient was bright, alert, and responsive. Pale-cyanosis was observed on the visible mucous membranes, Capillary refilling time of the mucous membranes was prolonged more than 2 sec. Abdominal palpation revealed no abnormalities. Thoracic auscultation heard a grade IV/VI systolic murmur at the right sternum and a grade III/VI systolic murmur at the left basal area. Systolic blood pressure and pulse rate were 120 mmHg (measured by the Doppler method) and 140 beats/min, respectively. Laboratory tests including complete blood count and blood chemistry found no significant abnormalities except for mild anemia packed cell volume was 32.7% (RR: 37% to 55%)]. Molecular detections for *Babesia canis, Ehrlichia canis, Hepatozoon canis* using PCR and *Anaplasma* spp. using Realtime-PCR were not found. The diagnosis was based on signalment, clinical signs, and physical

examination including thoracic auscultation, thoracic radiographs, and echocardiography. For the echocardiography was performed with a Hitachi Arietta 65 ultrasound machine with a Hitachi S31 pediatric phased array – Arietta transducer (2–9 MHz). Patients were examined in a lateral recumbence position, from both the right and the left side on a cardiac table to acquire standard echocardiographic views.

Results and Discussion

Chest radiography in a right lateral view showed normal contours of trachea, heart and mediastinum (vertebral heart score 9) (Fig. 1) and therefore, echocardiography was subsequently performed.

Pertinent findings upon two-dimensional echocardiography in the right parasternal long-axis LV outflow tract view were a normal left ventricle and left atrium. The measurements of the right ventricular (RV) free wall, the ventricular septum and the left ventricular (LV) free wall were 11.5, 7.5 and 5.9 mm, respectively. These measurements indicate hypertrophy of the RV, because the normal thickness of RV free wall is generally less than half that of the LV free wall (Boon, 2002). A large ventricular septal defect (VSD) was located at the perimembranous region, directly under the aortic valve, and overriding of the aorta was observed (Fig. 2A). Shunting of the blood flow from the LV to the RV through the VSD is always present during systole (Fig. 3). In the view of the right parasternal long-axis four-chamber, showed right atrium enlargement [23.5 mm; (RR: 12.2 ± 1.9 mm)] (Boon, 2002) (Fig. 2B). Right parasternal short axis view at aorta-main pulmonary artery level and continuous wave Doppler showing severe valvular pulmonic stenosis (PS). The pressure gradient of the right ventricular outflow tract at the pulmonic valve was approximately 91 mmHg. (Fig. 4). Several differential diagnoses are possible for exercise intolerance, cyanosis, and a grade IV/VI systolic murmur. However, considering the echocardiographic findings, the only possible diagnosis, in this case, was TOF without secondary polycythemia. The lack of polycythemia suggests that the patient did not have a large right-to-left shunt.

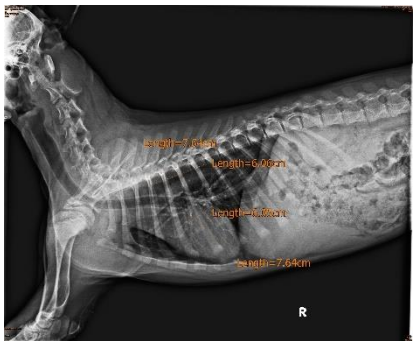

Figure 1 Right lateral thoracic radiographs. This figure shows normal heart and mediastinal contours (vertebral heart score 9).

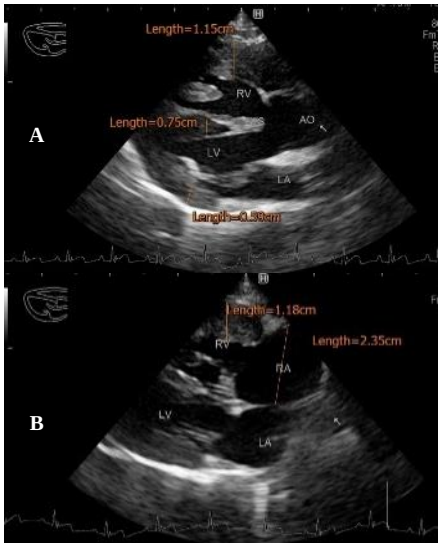

Figure 2A Right parasternal long axis 5-ch view, shows a right ventricular hypertrophy (two times the thickness of the left ventricular free wall), displacement of the aortic root on the right side, over the interventricular septum (overriding aorta).

Figure 2B Right parasternal long axis 4-ch view, shows a right atrium was enlarged (23.5 mm) and right ventricular hypertrophy. Aorta (AO), Interventricular septum (IVS), right atrium (RA), right ventricle (RV), left atrium (LA), left ventricle (LV).

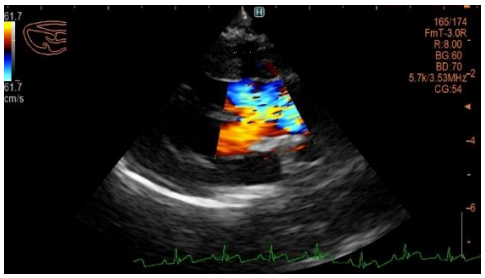

Figure 3 Color Doppler when applied at the right parasternal long axis left ventricular outflow tract (LVOT) view, showed turbulent blood in systole at the shunt.

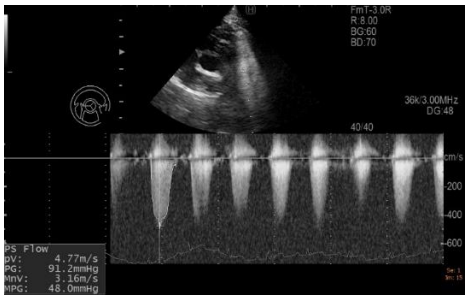

Figure 4 Spectral continuous wave Doppler study of main pulmonary artery with severe pulmonic stenosis (PS). There is an increased velocity of the ejection flow, which is 4.77 m/s (estimated right ventricle to pulmonary artery pressure gradient of 91 mmHg)

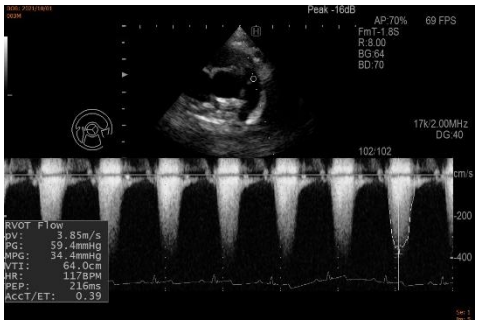

Figure 5 Spectral continuous wave Doppler study of main pulmonary artery after 1 month of treatment with atenolol. The ejection flow was 3.85 m/s (estimated right ventricle to pulmonary artery pressure gradient of 59 mmHg)

Due to financial reasons, owners declined the CT scan and the patient was medicated with atenolol 0.3 mg/kg SID PO for severe pulmonic stenosis. During the following month (1 month), the patient's clinical

condition was stable. The echocardiographic values showed improvement. (PS maximal velocity 3.85 m/s; PG 59 mmHg) (Fig. 5). The patient's quality of life on the day of examination remained good. In conclusion, this report describes a first case of TOF in an Alaskan malamute in Thailand.

The degree of right to left shunting and cyanosis depends on the severity of the pulmonic stenosis and the difference between systemic vascular and pulmonary vascular resistance. Any activity increases the right to left shunt. Oxygen saturation in the right atrium, right ventricle, pulmonary artery, and left atrium is normal; it is reduced in the left ventricle and aorta (due to the right to left shunt). The prognosis for long-term survival was poor. The usual sequelae include progressive polycythemia and progressive cyanosis. Dogs and cats rarely survive more than 3-4 years.

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