Extreme Tetralogy of Fallot in a Dog

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ABSTRACT. A four-month-old female Labrador retriever was brought to the Tokyo University of Agriculture and Technology Animal Medical Center for examination of its main symptoms of cough, tachypnea and exercise intolerance. Upon examination, the dog was found to have cyanosis and inadequate growth. Echocardiography revealed tetralogy of Fallot. Cardiac catheterization confirmed that the main pulmonary artery was completely occluded and that blood flowed from the aorta to the pulmonary artery. Accordingly, the animal was diagnosed with extreme tetralogy of Fallot.

KEYWORDS: canine, cardiac catheterization, extreme tetralogy of Fallot.


Tetralogy of Fallot (TOF) is a congenital heart disease characterized by four pathological abnormalities of morphology: ventricular septal defect (VSD), aortic overriding, right ventricular hypertrophy and pulmonary stenosis. Its incidence in dogs has been reported to be 0.0025% [6]. TOF with complete occlusion of the pulmonary artery is termed “extreme TOF”, and it is a more serious condition than regular TOF. To the best of our knowledge, there has been no previous report of extreme TOF in a dog. This report describes a canine case of extreme TOF.

A four-month-old female Labrador retriever was brought to the Tokyo University of Agriculture and Technology Animal Medical Center for thorough examination of cough, tachypnea and exercise intolerance.

Auscultation revealed a heart rate of 120 beats per min and a prolonged first heart sound (S1). The animal was small and thin compared with her littermates, with a body weight of 11.8 kg (body condition score 2/5). Capillary refill time was extended to more than 2 sec, and cyanosis was observed on the visible mucous membranes. The complete blood count and biochemical findings were within the normal ranges. According to oscillometric blood-pressure measurements, the systolic blood pressure was 136 mmHg, the diastolic blood pressure was 57 mmHg and the pulse pressure was 79 mmHg. Despite the animal was given 100% O2, oxygen saturation of the arterial blood (S\text{O}_2), measured by a pulse oximeter at the ear pinna, was only 94%.

Electrocardiography showed the presence of a right-axis deviation (−158°) as well as a deep S-wave (−1.8 mV) and a QRS width of 71 ms in lead II. These findings suggested that the right ventricle (RV) was enlarged [14].

Chest radiography in a lateral view showed an increased radiopacity of the aortic arch and an enlargement of the RV (vertebral heart score 10.9) [9] (Fig. 1). Furthermore, enlargement of the right atrium and the RV was indicated in the dorsal–ventral view (cardiac thorax ratio 77%) [9] (Fig. 2).

In the right parasternal long-axis four-chamber view of the echocardiogram, the measurements of the RV free wall, the ventricular septum and the left ventricular (LV) free wall were 11.7, 11.5 and 9.0 mm, respectively. These measurements indicate hypertrophy of the RV, because the free wall thickness of a normal RV is generally less than half that of the LV [2]. In the view of the LV outflow tract, the presence of a 9.5-mm ventricular septum defect (VSD) and overriding of the aorta were observed (Fig. 3). Shunting of the blood flow from the RV to the aorta through the VSD was confirmed by means of color-flow Doppler analysis. The cardiac index (cardiac output to body surface area ratio) was 2.7 l/min/m²: the body surface area was based on the body weight [4], and the cardiac output was calculated from the waveform for the aortic blood flow. The presence of a VSD could also be seen in the right parasternal short-axis view at the level of the aortic valve (Fig. 4). The main pulmonary artery did not give a distinct image as a result of an infundibular septal deviation, as indicated in the high-echo area (Fig. 4). In the same view, pulse Doppler analysis and color-flow Doppler analysis were unable to confirm the presence of blood flow.
from the RV to the pulmonary artery.

Next, a cardioangiography was performed. For the right side of the heart, an 80-cm long catheter with an external diameter of 4 French (1.35 mm) of the multipurpose Cournand type (TCA7118015; Technowood, Tokyo, Japan) was used. For the left side of the heart, a 110 cm-long catheter with an external diameter of 6 French (1.8 mm) of the angiographic catheter type (RH-6SP0061i; Terumo, Tokyo, Japan) was used. Atropine sulfate (Atropine Sulfate Injection 0.5 mg; Mitsubishi Tanabe Pharma Corp., Osaka, Japan) at a dosage of 0.05 mg/kg was administered subcutaneously as a pre-anesthetic medication for the cardioangiography. After 10 min, 4 mg/kg of propofol (Rapinovet; Intervet K.K., Tokyo, Japan) was administered intravenously to induce anesthesia. Subsequently, a tracheal tube was inserted, and inhalation anesthesia with isoflurane (Isoful; Dainippon Sumitomo Pharma Co., Ltd., Tokyo, Japan) under positive-pressure ventilation was applied. Angiocardiography was performed by using 1.4 ml/kg (iodine concentration 300 mg/ml) of a contrast medium (Oyparomin 300; Fuji Pharma Co., Ltd., Tokyo, Japan). The skin on the left side of the neck was cut open, and the left cervical vein and left cervical artery were identified. Subsequently, surgical threads were used to hold the blood vessel, and small cuts were made in the walls.
of the cervical vein and the cervical artery. Hemorrhage from these vessels was controlled by means of the surgical threads. Catheters were passed through the cut in the side of the left cervical vein to the right chambers of the heart and through the cut in the side of the left cervical artery to the left chambers of the heart. Selective RV angiography revealed that contrast medium flowed directly into the aorta from the RV instead of entering the pulmonary artery. Furthermore, contrast medium flowed from the aorta to the pulmonary artery through an abnormal blood vessel at a position distal to the regular opening of the ductus arteriosus (Fig. 5). Direct blood pressure measurement with a cardiac catheter indicated a systolic pressure of 75 mmHg in both the RV and LV, showing that the pressure was equal in the two ventricles. An analysis of the oxygen partial pressure was also performed simultaneously. The oxygen partial pressures in the LV, RV and aorta were 111, 52 and 75 mmHg, respectively.

On the basis of the above test results, the animal was diagnosed as having extreme TOF. The animal was administered the following oral medications: 0.01 mg/kg of digoxin (Digoxin-KY tab; Nippon Pharmacy, Tokyo, Japan) once a day, 1.5 mg/kg of alacepril (APINAC; DS Pharm Animal Health Co., Ltd., Osaka, Japan) once a day and 10 mg/kg of dipyridamole (Persantin-L Capsules; Boehringer Ingelheim Co., Ltd., Tokyo, Japan) twice a day.

The animal remained alive 30 months after its first visit to the animal medical center. At this time, its cardiac index was 7.8 l/min/m², and its $S\text{O}_2$ was 95% with the inhalation of 100% $O_2$. Both values were improved from those observed during the initial examination. While coughing occasionally occurred and cyanosis was observed during states of excitement, the animal’s exercise intolerance and respiratory state had improved as it had grown. Because of the excellent appetite of the animal, its body weight had increased to 16.4 kg. Furthermore, radiography and blood tests did not detect any clear progression of the disease. We are therefore continuing to administer medical therapy with follow-up observation.

Extreme TOF is a rare congenital heart disease, and only veterinary cases have been reported in cattle [8]. One of the characteristics of TOF is stenosis of the RV outflow tract. In extreme TOF, the stenosis becomes more serious, resulting in occlusion of the pulmonary artery [15]. In this condition, blood for gas exchange in the lungs is supplied through a patent ductus arteriosus or a major aortopulmonary collateral artery [1, 5, 11, 15]. In three bovine cases of extreme TOF reported by Nakada et al., blood was carried to the lung through the arterial duct [8]. However, the numbers of arterial ducts, the positions of the arterial ducts and the defects or occlusions of the main pulmonary artery, and other concomitant cardiac malformation differed from animal to animal. Furthermore, the main pulmonary artery was completely absent in two of the cases [8]. In the case of our dog, the presence of the main pulmonary artery was not clear, and that of a single arterial duct was demonstrated by echocardiography and by cardioangiography. On the basis of these findings, we consider the heart abnormalities in our case to resemble those in Case 2 reported by Nakada et al. (pulmonary artery completely absent with a left arterial duct). At present, it can be considered that extreme TOF is a category of congenital heart disease called “pulmonary atresia with VSD” (PA-VSD) [10]. In extreme TOF, the pulmonary artery is closed by a critical pulmonic stenosis and, although its form varies, a portion of the main pulmonary artery exists [13]. In contrast, it is more suitable to apply the term PA-VSD when the main pulmonary artery is anatomically absent [7, 10]. However, as the animal in this case remained alive, we were unable to perform a detailed anatomical evaluation of the main pulmonary artery and the abnormal ducts that connected the aorta and the pulmonary artery.

In the present case, the existence of an occlusion of the pulmonary artery was confirmed by echocardiography, although the presence of an abnormal vessel between the aorta and the pulmonary artery was not confirmed. Possible reasons for this observation are that the abnormal vessel existed distally to the usual position of the opening of the arterial duct, and that blood flowing from the aorta was directed downstream of the pulmonary artery, as the pulmonary artery was occluded [8, 12]. Furthermore, the position and form of the heart in the thoracic cavity in our case may have differed from that in normal dogs. This might explain why the abnormal vessel could not be visualized on the cross-section of transthoracic echocardiography. However, the abnormal vessel could be visualized by cardioangiography. Accordingly, extreme TOF might be accurately diagnosed by applying a combination of several tests. In human medicine, magnetic resonance imaging (MRI) is considered effective for diag-

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**Fig. 5.** Selective right ventricular angiography. The catheter tip was located at the right ventricular apex. Most of the blood ran directly into the aorta from the right ventricle instead of entering the pulmonary artery. Also, the blood flow ran partially from the side of the aorta to the side of the pulmonary artery through an abnormal blood vessel at a position distal to the regular opening of the ductus arteriosus (arrow). Ao: aorta, LA: left atrium, RA: right atrium, RV: right ventricle, PA: pulmonary artery.
nosing extreme TOF [1]. Therefore, in the future, it may be possible to identify the type of abnormal vessel present by performing MRI while the animal is still alive.

The Rastelli procedure is recommended as a radical treatment for extreme TOF. In this procedure, the RV and main pulmonary artery are connected with a valved graft, while the VSD is closed [7, 13, 15]. At present, the animal in our case is being closely monitored, and several test results have clearly indicated that there is no progression of the disease. The animal’s general condition has gradually improved with growth. In cases where a shunt vessel is present, it has been reported that the type of defect influences the prognosis and the survival time in extreme TOF [3, 7, 11]. The animal’s hemodynamic status has been improved by the presence of adequate pulmonary blood flow volume through the shunt vessel, as well as by the medical therapy, producing positive effects on its prognosis. However, if the animal’s condition worsens in the future, the Rastelli procedure may have to be considered.

REFERENCES


