A retrospective study of tetralogy of Fallot in dogs

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Abstract: Tetralogy of Fallot is a rare congenital heart defect; however, it is the most common cyanotic heart malformation in humans and animals. It consists of a ventricular septal defect, pulmonic stenosis, displacement of the aortic root, and hypertrophy of the right ventricle. At the Department of Internal Medicine Clinic of Diseases of Horses, Dogs, and Cats at the Wroclaw University of Environmental and Life Sciences, patients with Fallot syndrome constituted 4.44% of all patients with congenital heart diseases. There was no breed predisposition found. The most common clinical signs were exercise intolerance, dyspnea, and cyanosis of the mucous membranes, but it can also be asymptomatic. Echocardiography is considered to be the best method for diagnosing this congenital heart disease. Contrast angiography can be used to confirm the diagnosis or as a complement to diagnosis. Surgery is the only effective method of treatment; however, for animals, we only provide palliative treatment.

Keywords: Tetralogy of Fallot, cyanosis, congenital heart disease

1. Introduction

The anatomic features of tetralogy of Fallot (ToF) were first described by the Danish anatomist Nils Stensen in 1673, but in 1888 the French doctor Etienne Fallot gave a detailed anatomical and clinical description of this heart defect (1). ToF is a complex congenital heart disease that consists of 4 components: 1) ventricular septal defect (VSD); 2) stenosis of the right ventricular outflow track (RVOT stenosis) and pulmonic stenosis (PS); 3) displacement of the aortic root on the right side, over the interventricular septum, a so-called rider aorta; and 4) hypertrophy of the right ventricle. If only RVOT stenosis, right ventricular enlargement, and an atrial septal defect are present, it is called trilogy of Fallot, but 10%–15% of ToF cases are accompanied by atrial septal defects such as patent foramen ovale; such cases are called pentalogy of Fallot. ToF has been described in animals such as dogs, cats, horses, sheep, pigs, cattle (2), and bears (3). In humans, ToF makes up about 10% of all congenital heart diseases and is the most recognized cyanotic defect after the first year of life (4). ToF is characterized by high variability and a variety of anatomic changes. VSD is usually a large perimembranous defect that can also take up most of the septum or be located beneath the pulmonary valve (5–7). The pulmonary valve is often stenotic; it can be bicuspid or dysplastic (so-called unicuspid), or it can have deformed cusps as fibrous nodules. The cusps are usually thickened and shortened. The pulmonary trunk has an abnormally small diameter and is hypoplastic. PS can exist as an isolated subvalvular stenosis, a combination of subvalvular and valvular stenosis, or, rarely, a simple valvar stenosis (8).

RVOT stenosis causes hypoperfusion of the lungs, resulting in reduced blood oxygenation. High pressure in the right ventricle causes a right-to-left shunt, which means that unoxygenated blood from the right ventricle gets into the left ventricle via the VSD before returning to systemic circulation. The development of hypertrophy of the right ventricle is a result of pressure overload, secondary to stenosis of the pulmonary outflow tract. As a result, unoxygenated blood is delivered to the tissues, which causes cyanosis. ToF is the most common cyanotic heart defect diagnosed in humans (9) and animals (10).

2. Materials and methods

At the Clinic of Diseases of Horses, Dogs, and Cats of the Department of Internal Medicine at the Wroclaw University of Environmental and Life Sciences, 225 dogs were diagnosed with congenital heart disease. Among these dogs, there were 10 dogs with Fallot syndrome.
Diagnosis was based on clinical signs and physical examination including auscultation, electrocardiography, echocardiography, and ventriculography. A postmortem examination was also obtained (Figure 1). For the electrocardiographic examinations, a BTL-08 SD machine with bipolar and unipolar standard leads and crocodile clips was used. The echocardiography was performed with an Aloka Prosound 4000+ ultrasound machine with a transducer (3.5–8 mHz). Patients were examined in a standing position, from both the left and the right side. Ventriculography was performed with a fluoroscopy system to confirm the diagnosis from the echocardiographic examination, or as a complement to this method.

3. Results
There were 10 dogs diagnosed with Fallot syndrome, 8 with tetralogy of Fallot, and 2 with pentalogy of Fallot. This number constituted 4.44% of all patients with congenital heart diseases. Among these 10 dogs, there were 4 times as many males as females (8:2). In our clinic, Fallot syndrome was diagnosed in a single dog of each of the following breeds: Dachshund, Shiba Inu, Bernese mountain dog, Bavarian mountain hound, Beagle, Airedale terrier, Havanese, and one mixed breed. French bulldog was the only breed in which 2 unrelated dogs had ToF, but there was no breed predisposition found.

In our dogs, symptoms overlapped with those described in the literature (11), namely exercise intolerance, cyanosis, and dyspnea. Blood tests (complete blood cell count, alkaline phosphatase, alanine transferase, urea, and creatine) did not show any significant changes. A systolic murmur was heard at the base of the heart. On the left side, the murmur of the pulmonary artery was dominant. Usually, the murmur was very loud, but this generally depended on the degree of pulmonary stenosis. The electrocardiogram examination showed a physiological sinus rhythm in most of the cases, but there was sinus tachycardia only in the advanced stages. Hypertrophy of the right ventricular wall caused an increase in the amplitude of the S wave in leads I, II, and III. Often there was an apparent deviation of the heart axis to the right (>100°). Echocardiographic examination usually provided a final diagnosis. In dogs with Fallot syndrome, hypertrophy of the right ventricular free wall was evident. In most cases, the thickness of the right ventricular wall was even greater than the thickness of the left ventricular wall. In the apical 4-chamber view, the VSD and “rider aorta” were visible (Figure 2), along with the atrial septum defect in the case of pentalogy of Fallot. The flow between the ventricles was visualized using Doppler ultrasound (color and spectral). The velocity of the pulmonary flow (measured from the right side from a short-axis, parasternal view) was often higher than normal due to stenosis. In dogs that were also examined by ventriculography, during selective fluoroscopic examination, the contrast medium was injected through a catheter into the great vessels and the chambers of the heart. This made the pathological flow, shunts, and stenosis visible. In dogs with ToF, after introduction of the angiographic catheter into the external jugular vein and injection of contrast medium into the right ventricle, the hypoplastic pulmonary trunk with pulmonary valve stenosis, hypertrophied right ventricle, pathological right-to-left shunt via VSD to the aortic root, and occasionally aortic valve regurgitation were all visible (Figure 3).

![Figure 1](image1.png)

**Figure 1.** Necroscopic examination. The green arrow indicates a hypertrophy of the right ventricular wall and the blue arrow points to a VSD.

![Figure 2](image2.png)

**Figure 2.** Echocardiographic examination. The pathological right-to-left shunt via VSD is visible as a turbulent, colorful flow (blue arrow). LA = left atrium, LV = left ventricle, RA = right atrium, RV = right ventricle, and VSD = ventricular septal defect.
4. Discussion

The prevalence of ToF in humans ranges from 3% to 10% of all congenital malformations (9,12,13). In dogs, the reported occurrence of ToF is rarer and ranges from 0.6% to 7% (11,14–17). These results are predominantly for tetralogy of Fallot and there is no mention of other forms of this syndrome. In our study, all forms of Fallot syndrome were considered, but even if we consider only tetralogy, its occurrence is average. There was no sex predisposition reported in the previous studies. Despite the apparent predominance of males in our study, we cannot assume that there is such a predisposition because of the low number of examined dogs. Among the dogs that we examined, there seems to be no breed predisposition. The breeds in which Fallot syndrome was diagnosed in our clinic are different from those described in the available literature.

Echocardiography is considered to be the best method for diagnosis of this congenital heart disease. To confirm diagnosis, or as a complement to this method, contrast angiography can be used.

In humans, a few genes have been identified in ToF etiology, including Jagged-1 (18), NKX2-5 (19), and ZFPM2 (zinc finger protein multitype 2) (20). In dogs, genetic research is just beginning, and in Keeshond dogs, autosomal recessive inheritance and a possible genetic etiology have been observed (11,21,22), but no genetic tests were performed in this study.

Surgery is the only effective method of treatment. A complete correction of Fallot syndrome requires many surgeries using cardiopulmonary bypass. For this reason, there are only palliative procedures in animals. Pulmonary flow can be increased by surgical formation of the systemic-pulmonary anastomoses using 2 main techniques: 1) a "side-by-side" type of anastomosis between the ascending aorta and the pulmonary artery, or 2) anastomosis of the subclavian artery to the pulmonary artery (10). Pharmacological treatment is limited to diuretics such as loop diuretics, beta-blockers, and ACE-I, which may reduce clinical signs. Drugs that increase blood pressure should be avoided, and a low sodium diet is recommended. Oxygen therapy, puncture, and removal of fluids from the pleural and abdominal cavity may be needed in case of deterioration of the general condition. In our clinic, we recommend only standard palliative pharmacological therapy or euthanasia.

Congenital heart malformations like Fallot syndrome are rare, but the development of diagnostic methods facilitates their recognition and leads us to conclude that they are more common than we used to think.

Figure 3. Selective lateral right ventricular angiocardiogram. The white arrow indicates the catheter, blue = brachiocephalic artery, burgundy = aorta, red = aortic root, brown = coronary artery, yellow = pulmonary artery, pink = aortic valve regurgitation, and green = right-to-left shunt via VSD.

References


