

Tetralogy of Fallot

Tetralogy of Fallot is a rare congenital cardiac defect diagnosed in dogs and cats. It is characterized by several congenital cardiac defects. The Keeshond breed is predisposed, and English Bulldogs have been over-represented, but any breed can be at risk. It is diagnosed via echocardiography – it is a challenging diagnosis and best evaluated by a veterinary cardiologist.

The defects which make up tetralogy include valvular pulmonic stenosis, a ventricular septal defect, over-riding (dextropositioned) aorta and right ventricular concentric hypertrophy (thickening). Pulmonic stenosis refers to malformation/dysplasia of the pulmonic valve where it is not opening normally – this leads to obstruction to blood leaving the right ventricle causing pressure overload. This increased work that the right ventricle must perform to eject blood across a narrowed opening leads to the right ventricular concentric hypertrophy. Some animals with Tetralogy will have hypoplasia of the pulmonary artery as well which further limits flow from the right ventricle. A ventricular septal defect (VSD) is a hole present in the muscular wall that separates the right and left ventricle. Finally, an over-riding aorta refers to a malorientation of the aorta where the aorta is positioned over both the right and left ventricle (the aorta should be positioned only over the left ventricle).

The consequence of a tetralogy of Fallot is shunting of deoxygenated blood into the systemic circulation across the VSD into the dextropositioned aorta. This leads to peripheral cyanosis and hypoxia. Affected animals will develop a condition called polycythemia over time as a result of hypoxia – this is where there is excessive red blood cell production to compensate for the hypoxia. Affected animals will have exercise intolerance, trouble breathing, cyanotic mucous membranes, weakness and collapse.

There is no definitive treatment for this condition. Surgeries have been attempted in animals with mixed results. Medical management may provide some relief for some animals – this may entail the use of a beta blocker called propranolol which may limit the degree of right to left shunting. Treatment of the polycythemia may include periodic phlebotomies (a procedure to remove a certain volume of blood from the body and replace with fluids to 'dilute' the blood) and a medication called hydroxyurea. Hydroxyurea is a chemotherapy drug which affects the bone marrow production of red blood cells.

Prognosis is guarded – smaller dogs and cats may tolerate their defect in the long term better than larger animals. Sudden death has been reported in patients with Tetralogy.

