Pathoanatomy

The primary pathoanatomy seen in dogs with Tetralogy of Fallot (ventricular septal defect, pulmonic stenosis and overriding aorta) can all be explained by embryological developmental defects of the endocardial cushions comprising the conotruncal septum. (Patterson and others 1974) This is because the sinistroventral conus cushion and sinistrolateral truncus cushion give rise to the left part of the ventricular septum, and the left pulmonary valve leaflet respectively. The dextrodorsal conus cushion and dextrosuperior truncus cushions give rise to the right portion of the ventricular septum and the right pulmonary valve leaflet respectively. Finally, the anterior valve leaflet forms from the intercalated valve cushion. Abnormalities of conotruncal septal development, therefore, give rise to a spectrum of intracardiac defects the most severe viable form of which is Tetralogy of Fallot. (Patterson 1989) The degree of ventricular hypertrophy that accompanies the primary abnormalities will depend on the severity of the pulmonic stenosis and the size of the septal defect. The right ventricular outflow tract obstruction forms the progressive component of this disease because of the contribution of secondary infundibular muscular hypertrophy.

Pathophysiology

Right to left shunting of blood results in reduced pulmonary arterial blood flow and admixture of “right heart” and “left heart” blood; resulting in systemic hypoxia and cyanosis. Compensatory polycythemia is usually seen as the disease progresses. The contribution of infundibular muscle to the right ventricular outflow tract obstruction can create both a dynamic and progressive component to this disease, accounting for worsening of signs with exercise and progression of disease despite fixed valvular and septal abnormalities.

Therapeutic options

Medical/conservative: Some mildly affected animals not require treatment and will still enjoy a good quality of life; others may require medical therapy (in the form of beta-blockers) to give an acceptable quality of life. It has been suggested that approximately 25% of affected animals will not be controlled by conservative/medical means (Eyster and others 1976).

Surgical: Surgical treatments fall into two main categories: primary repair and palliative procedures.

Primary repair of tetralogy of Fallot has been described several times in the veterinary literature (Heritage and others 1983, Lew and others 1989) this requires cardiopulmonary bypass, advanced surgical expertise and currently carries a high mortality rate.
**Palliative surgical procedures:** These were used extensively in human medicine prior to the advent of cardiopulmonary bypass and primary repair. Currently, some are used in infants as a “bridge” to primary repair, in severely affected individuals.

1. **Potts anastomosis.** (Side to side anastomosis of the aorta to the left main pulmonary artery) This is technically demanding and requires special instrumentation. It has been described in a small group of dogs (Ringwald and Bonagura 1987). Disadvantages of this procedure include the “high price” paid if the aorta tears during or after the procedure and the fact that flow across the anastomosis depends on the size of the aperture created. It is possible to “overdo” things and create left-heart overload by increasing the flow to the pulmonary artery too much.

2. **Blalock anastomosis.** (Anastomosis of the left subclavian artery to the left main pulmonary artery) The advantage of this procedure is that flow rate is dictated by the size of the subclavian and, therefore, pulmonary overload is very unlikely. “Kinking” of the vessel at the junction with the aorta, as it turned 180° to join the pulmonary artery seems to have been a major problem (Weber and others 1995, Eyster and others 1977). This problem led to the modification of the procedure to include resection of a diamond-shaped piece of tissue from the left subclavian:aortic junction. (making it a lot more risky – authors opinion). Eyster (1977) concluded that the Blalock anastomosis worked “well” in dogs over 10kg bodyweight.

3. **Modified Blalock-Taussig (mBT) anastomosis.** The development of synthetic materials such as polytetrafluorethylene (PTFE) (Gore-Tex) that is relatively non-thrombogenic and can be created in tube form, has allowed surgeons to modify existing techniques. This has been used to good effect to overcome some of the complications associated with using local native tissues for the Blalock anastomosis (Kay and others1983). The use of PTFE to create a modified Blalock-Taussig anastomosis has been reported in a dog that failed conventional Blalock anastomosis (Weber and others). In this operation, the shunt was sutured from the main brachycephalic trunk to the pulmonary artery (because the left subclavian had already been sacrificed). The author has used a PTFE shunt from the left subclavian to the pulmonary artery in dogs with cyanotic, polycythemic tetralogy, to provide long-term palliation of clinical signs (Brockman and others 2007). A mBT shunt can also be created using native tissues such as the jugular vein (Pelosi and others 2006).

4. **Others:** Microvascular anastomosis of the left internal thoracic artery to the pulmonary artery has been described in a cat that had a failed “Fontan” procedure (anastomosis of the right atrium to the pulmonary artery) (Miller and others 1985).

**Outcome:** Long-term palliation can be achieved for dogs with ToF, although there is a sense that larger dogs and dogs that are older at the time of presentation have a more durable palliation, the number of dogs treated by this operation remains so small that firm conclusions cannot yet be made.
References: