Congenital Heart Disease in Dogs

Congenital heart disease is the term used to describe heart disease that is present from birth. Some types of congenital heart disease in certain breeds have been shown to be heritable, or genetically passed down from the parents to the puppy.

A dog's heart is anatomically similar to a human heart. There are four chambers, two on the right side, and two on the left. The upper chambers are called atria, and the lower chambers are called ventricles. In a normal functioning heart, blood enters the right atrium from the head, neck and abdomen and flows through the tricuspid valve to the right ventricle. From there it is pumped through the pulmonary valve into the pulmonary artery, then into the lungs where it receives oxygen. Blood re-enters the heart via the left atrium and travels through the mitral valve into the left ventricle. Finally, oxygenated blood exits the heart through the aortic valve into the aorta.

A congenital heart defect, or combination of defects, can occur as a malformation of any valve, chamber, or great vessel. A defect can also involve abnormal connections between the heart chambers. The most common congenital heart diseases in dogs include patent ductus arteriosus (PDA), pulmonic stenosis (PS), and subaortic stenosis (SAS). Because these are all potentially inherited defects, it is recommended that affected dogs and their parents be removed from breeding programs.

A PDA is caused by the lack of closure of the ductus arteriosus, a blood vessel connecting the aorta and pulmonary artery. This vessel is open during fetal development to allow blood to bypass the developing (nonfunctional) lungs in the fetus. At birth as the puppy breathes and the lungs are inflated, the ductus should close and blood should begin to flow in a normal pattern. An open, or patent

ductus arteriosus, is a congenital defect where blood continues to be shunted between the pulmonary artery and the aorta (usually from the aorta to the pulmonary artery, or left-to-right shunting). The result is a blood volume overload in the pulmonary arteries and veins and left side of the heart. Long standing volume overload results in dilatation of the left cardiac chambers and left-sided congestive heart failure (fluid in the lungs). Many patients do not survive beyond a year of age if this defect is not corrected.

PS is a narrowing of the pulmonic valve opening resulting from abnormal formation of the valve. The right ventricle has to pump blood out of a narrowed opening. This requires more work, and higher pressures must be achieved to push the blood out of the smaller opening. The right ventricle becomes enlarged, and in some patients, right-sided heart failure (fluid in the abdomen and an enlarged liver) results. Depending on the degree of severity, some dogs live normal life spans without medication. More severely affected dogs can succumb to right-sided heart failure within the first few months to years of life if left uncorrected. PS is most commonly found in small breed dogs.

SAS is a narrowing found just below the aortic valve and is most often the result of fibrous nodules or a fibrous ring of tissue that develops within the first 4-8 weeks of life in the left ventricular outflow tract. The resulting murmur often intensifies over the first year of life. Because the flow of blood is obstructed from the left ventricle through the aorta, the left ventricle must work harder and at higher pressures to eject blood. As a result, the left ventricle becomes enlarged, and in some patients, left-sided heart failure (fluid in the lungs) results. The thickening of the left ventricle and high pressures can also predispose these patients to serious arrhythmias (irregular heartbeats). Again, mildly affected dogs can live a normal life span. SAS most commonly affects large breed dogs.

Diagnosis

Most puppies diagnosed with congenital heart disease are asymptomatic when

they are first diagnosed. Congenital heart disease in dogs is typically first identified when a murmur is noted on physical examination during one of the first routine vaccination visits. A murmur is turbulent blood flow that creates vibrations which can be heard when a stethoscope is used to listen to the heart sounds. Innocent and physiologic murmurs are quiet, not associated with a heart defect, and commonly resolve by 16 weeks of age. Congenital heart defects typically produce loud murmurs that persist or become louder over time. A PDA causes a continuous (washing-machine) type murmur, whereas PS and SAS cause an ejection murmur heard when the heart is pumping blood forward to the body and lungs.

Referral to a veterinary cardiologist is recommended for a complete echocardiogram (ultrasound of the heart) to evaluate the cardiac anatomy and function of the heart and great vessels. Color flow and spectral Doppler echocardiography are essential to the congenital examination to identify the abnormality in blood flow, and classify the severity of the defect. The majority of congenital disease can be accurately diagnosed with echocardiography alone. In very complex congenital defects, a cardiac catheterization and angiogram procedure may be necessary to fully determine the extent of the defects. Additional diagnostics such as thoracic radiographs, blood pressure and blood work may be recommended.

Complications

Over time, typically within the first year of life, a PDA leads to volume overload of the lungs and left heart resulting in congestive heart failure (fluid in the lungs), and sometimes atrial fibrillation (a rapid irregular heart beat). This will cause symptoms such as cough, difficulty breathing, weakness and collapse. Left untreated these patients will succumb to their heart disease. Rarely, the PDA can have blood flow in the opposite direction (from the pulmonary artery to the aorta, or right-to-left shunting). In this case sufficient blood never reaches the lungs to pick up oxygen, and signs of cyanosis (blue gums), weakness, lethargy, intolerance to exercise and episodes of collapse can occur. Patients with right-to-

left shunting rarely live beyond 2-3 years of age, as surgical correction is not an option for these patients.
Patients with severe PS typically develop clinical signs by 2-3 years of age. Symptoms include weakness, lethargy, intolerance to exercise and episodes of collapse with exertion or excitement. More rarely, signs of right-sided heart failure (fluid in the abdomen and enlarged liver) can occur. Sudden death very rarely occurs in PS patients. Patients with mild to moderate PS may live a normal life span with good quality of life and little or no need for medication.

Patients with severe SAS typically develop clinical signs by 2-3 years of age. Symptoms of severe SAS include weakness, lethargy, intolerance to exercise, episodes of collapse and even sudden death due to the high risk of developing severe irregular heartbeats (ventricular arrhythmias). Less commonly, signs of left-sided heart failure (fluid in the lungs) can occur, producing signs of coughing and difficulty breathing. Any SAS patient is at increased risk for developing infections on the heart valves (vegetative endocarditis). This is a very serious complication that can result in heart failure or sudden death due to irregular heartbeats. Patients with mild SAS can live normal life spans with minimal medical therapy.

Treatment

Treatment options for PDA include closure of the open vessel by a transcatheter occlusion procedure or surgical ligation of the vessel. The less invasive option is transcatheter occlusion, or the placement of an occlusion device (an Amplatz® canine duct occluder) into the PDA through a catheter inserted into the femoral artery. This procedure is associated with a far more rapid recovery and less extensive hospital stay than a surgical (thoracotomy) repair. It still carries minor risks, including significant bleeding from the femoral artery, or puncture of the PDA with a catheter leading to fatal bleeding. Patients in heart failure at the time of diagnosis require routine heart failure treatment (medications) to stabilize them

prior to and often several months after definitive correction of the PDA by either of these two approaches.

Dogs with severe PS or clinical signs of right-sided heart failure (fluid in the abdomen and enlarged liver) often benefit from balloon valvuloplasty performed during cardiac catheterization. There are a few situations where correction is not an option, such as when an abnormal coronary artery is identified or if an extremely narrowed valve annulus (where the leaflets attach) is found. The balloon valvuloplasty procedure carries an approximate 80% success rate in reducing the severity of pulmonic stenosis to mild or moderate, thus reducing the risk of clinical signs and returning the expected life span towards normal. During this cardiac catheterization procedure, the right heart is accessed by way of the right jugular vein (right side of neck) and a balloon tipped catheter is strategically positioned across the narrowed valve under fluoroscopic guidance. Upon inflation of the balloon, the narrowed valve is snapped/popped/stretched open. Significant risks associated with this procedure are not common, but include fatal arrhythmias (abnormal heart rhythms) during the procedure and sudden death following the balloon inflation (related to collapse of the right ventricle secondary to the sudden reduction of pressure once the valves are opened further). Other mild risks associated with surgery include a small amount of bruising or swelling at the surgery site. Additionally, it is possible for the pulmonic valves to develop scar tissue and narrow again, requiring a second balloon procedure; although, this is very rare.

There are a few therapeutic options to correct SAS. Medical management with atenolol (a beta blocker) or sotalol (anti-arrhythmic and beta blocker) is usually recommended. The atenolol dosage must be gradually increased over time based on the dog’s tolerance of the medication and body weight. Should left-sided heart failure occur, routine congestive heart failure medications are used to improve the quality of life.

Prognosis

The majority of dogs with PDA, if left uncorrected, develop congestive heart failure and succumb to their disease by a year of age. If successfully corrected, these patients have an excellent chance of leading a normal life.
Dogs with mild to moderate PS do not typically have a reduction in their life span as a result of their heart disease and rarely show clinical signs. Dogs with severe PS, however, typically have a shortened life span and clinical signs of exercise intolerance, weakness and collapse and right-sided heart failure by 2-3 years of age, unless corrected by balloon valvuloplasty.

In general, we expect the severity of subaortic stenosis (SAS) to worsen/progress until the patient is close to mature body weight and then progress little after that time. All patients with SAS, regardless of the severity, have an increased risk of developing infections of their aortic valve (bacterial endocarditis). As such, appropriate precautions should be taken by administering antibiotics when there is an increased risk of bacteremia (bacteria in the bloodstream). This would include any surgical procedure, dental prophylaxis, fever or presence of a cut or wound. Severe SAS patients by 2-3 years of age will typically develop clinical signs of heart disease, which include exercise intolerance, collapse and even sudden death. Rarely left-sided congestive heart failure (fluid in the lungs) occurs. When this happens it can be managed medically for a period of time. Mild SAS does not typically affect the quality or quantity of a dog’s life.

Whenever a puppy is diagnosed with a heart murmur it can be devastating news. Our first goal is to determine what this will mean for the individual puppy and his or her new family. It is important to keep communication open with all involved. Discuss options for further diagnostics with your veterinarian and consider consultation with a veterinary cardiologist.

The sooner an accurate diagnosis can be made, the sooner a plan of action can be set in place to ensure the best outcome for your puppy. Our second goal is to educate all involved, so we can work toward preventing further breeding of dogs with heritable heart disease.

For more information on this disease, speak to the veterinarian who is treating your pet.