Introduction

Congenital heart disease makes up a small percentage of the cardiac disease we see in small animals. However, congenital heart diseases can lead to severe clinical signs and death at a young age if not identified and treated appropriately. Some of the congenital heart diseases seen in small animals can be improved, or even cured with catheter procedures or surgery. Therefore it is critical to recognize congenital heart disease and determine the appropriate treatment.

Innocent Murmurs

It is common for puppies and kittens to have soft heart murmurs at birth that resolve as the patient grows. Typically these murmurs are Grade I-II/VI in intensity and heard best at the left base. Innocent murmurs are often high-pitched or musical in character. Innocent murmurs usually resolve by 4 months of age. I will give a patient up to 6 months as long as there are no clinical signs and the murmur characteristics match with those of an innocent murmur. A patient with clinical signs of heart disease, a Grade III/VI or louder murmur, a continuous murmur, or a murmur heard best on the right side should be evaluated by a cardiologist as soon as possible.

Patent Ductus Arteriosus

Patent Ductus Arteriosus (PDA) is one of the most common congenital heart diseases seen in dogs. It is rarely identified in cats. PDA is seen most often in the Maltese, Poodle, Pomeranian, Collie, Bichon Frise, Yorkshire terrier, Chihuahua, and German shepherd breeds, although any breed can be affected. PDA is more common in females by a 2:1 to 3:1 ratio. With PDA, the ductus arteriosus remains open allowing blood to shunt from the aorta to the pulmonary artery. This leads to a volume overload of the left side of the heart and the lungs. Patients with a PDA typically have a loud, continuous murmur high on the left cranial thorax. In many cases there is a palpable thrill. The murmur of a PDA has been compared to machinery, a washing machine, and wind in a tunnel. In some dogs with a PDA, the stethoscope must be placed very far dorsal / cranial in the left axilla to detect the characteristic murmur. If left untreated, most dogs with a PDA will develop left sided congestive heart failure and die by 2 years of age. PDA can be corrected either by placement of an Amplatz ductal occluder via cardiac catheterization, or by thoracotomy and PDA ligation. Some dogs are not candidates for the ductal occluder based on their body size and the morphology of the ductus. Either method has a low incidence.
of complications and a high success rate when performed by experienced individuals. Following closure of the ductus, the left sided abnormalities seen on echocardiogram improve. The heart may not return to completely normal, but typically the dogs have a normal life span with no cardiac clinical signs.

Rarely we will see shunt reversal (right to left flow) in patients with large PDAs and secondary pulmonary hypertension. Dogs with this problem do not have the characteristic PDA murmur. These cases are managed with Sildenafil and if needed, treatment for polycythemia (phlebotomy, Hydroxyurea). Closure of the PDA is contraindicated.

**Pulmonic Stenosis**

Pulmonic Stenosis (PS) is also one of the more common congenital cardiac diseases in dogs. PS is rarely seen in cats. PS is seen most often in Chihuahuas, Labradors, Newfoundland, Beagles, Boxers, Bulldogs, and Terrier breeds. There are multiple forms of PS. The pulmonic valve may be thickened or the leaflets may be partially fused. In some cases, the entire pulmonary artery is too small. There may be abnormal tissue blocking blood flow below (subvalvular stenosis) or above (supravalvular stenosis) the pulmonic valve. In Boxers and Bulldogs, PS is often secondary to an abnormal coronary artery that encircles the right ventricular outflow tract. PS is broken down into mild, moderate, and severe categories based on the pressure gradient across the narrowed region. For dogs with moderate to severe stenosis, we will see secondary hypertrophy of the right ventricle and potentially right atrial enlargement. Dogs with PS are usually asymptomatic when the murmur is heard during initial puppy visits. Dogs with moderate to severe stenosis are at risk for exercise induced syncope or sudden death as they mature and exercise more intensely. The recommended treatment for moderate to severe PS is balloon valvuloplasty. The majority of patients respond well to the procedure, with an approximately 50% reduction in pressure gradient. There may be benefit to medical treatment with Atenolol, although this is not a substitute for the balloon valvuloplasty procedure. In Boxers or Bulldogs with an aberrant coronary artery, balloon valvuloplasty is contraindicated. Therefore it is critical to determine coronary anatomy prior to the balloon procedure in these breeds.

**Subaortic Stenosis**

Subaortic Stenosis (SAS) is one of the common congenital heart diseases seen in dogs. It is rarely seen in cats. SAS is most often identified in large breed dogs such as the Newfoundland, Golden Retriever, Boxer, Rottweiler, and German Shepherd. Even though SAS is considered a congenital defect, the lesion typically progresses as the patient reaches full adult size. In some cases of SAS, a murmur may not be present when the patient is under 12 weeks of age. The most common form of SAS is a fibrous ring encircling the left ventricular outflow tract. SAS can also be seen as a muscular protrusion from the high interventricular septum. Occasionally we can see valvular aortic stenosis or aortic hypoplasia.
Regardless of the form of SAS, the increased pressure load on the left ventricle leads to left ventricular hypertrophy if there is a significant obstruction present. Most dogs with SAS are identified by the presence of a loud murmur detected before clinical signs develop. The murmur is loudest at the left base, but may radiate strongly throughout the thorax and even up the carotid arteries. Dogs with moderate to severe SAS are at risk for exercise induced syncope or sudden death. Dogs with SAS are also at increased risk for aortic endocarditis because of chronic trauma to the aortic valve.

Dogs with moderate to severe SAS are often treated with Atenolol in hopes of slowing the heart rate, reducing any dynamic component to the obstruction, reducing the risk for arrhythmias, and reducing the risk for sudden death. The efficacy of this treatment has not been proven. Exercise restriction will also help reduce the risk for clinical signs.

Historically, balloon dilation of the stenotic region was attempted to for dogs with SAS. Although this lead to a reduction in the pressure gradient, there was no long term survival benefit. Recently, balloon dilation for SAS has been attempted again using a combination of a cutting balloon and a high pressure balloon. Short term results are encouraging, but it is too soon to tell if this procedure will lead to a long term survival benefit. Because of the increased risk for endocarditis, dogs with SAS receive prophylactic antibiotics prior to surgeries and dental procedures.

**Ventricular Septal Defect**

Ventricular Septal Defect (VSD) is occasionally seen in both cats and dogs. With a VSD, the interventricular septum does not form completely. The resulting defect is typically small and is located in the membranous septum near the aortic valve. Since the pressure is higher in the left side of the heart, blood will shunt from left to right across the VSD. The murmur of VSD is typically heard best on the right. With this defect, a louder murmur equates with a smaller defect and overall less serious condition. In most cases of VSD, there is either normal heart size or mild left heart enlargement. Patients with these echocardiographic findings typically have a normal life span with no cardiac clinical signs. There may be an increased risk of endocarditis, so prophylactic antibiotics are given for procedures. In patients with large VSDs, progressive left heart enlargement and congestive heart failure are possible. Medical treatment would be similar to other causes of CHF. In some cases, it may be possible to close the VSD with a device during cardiac catheterization. Rarely we will see shunt reversal (right to left flow) in patients with large VSDs and secondary pulmonary hypertension. These cases are managed with Sildenafil and if needed, treatment for polycythemia (phlebotomy, Hydroxyurea).
Mitral Valve Dysplasia

Mitral Valve Dysplasia is one of the more common congenital heart diseases in cats. MVD is also rarely seen in dogs. With MVD, malformation of the mitral valve and support structures leads to abnormal mitral valve function. Most feline patients will have systolic anterior motion of the mitral valve which causes a left ventricular outflow tract obstruction. If the obstruction is moderate to severe, secondary left ventricular hypertrophy will develop. Patients with MVD have a variable amount of mitral regurgitation. Mitral regurgitation and decreased left ventricular compliance will lead to left atrial enlargement and potentially congestive heart failure. Mitral stenosis is also possible, and can accelerate the development of clinical signs. Cats with MVD are often asymptomatic at diagnosis and are identified by a loud murmur in a young cat. There may be signs of exercise-induced syncope or respiratory distress if there is a significant left ventricular outflow tract obstruction. Treatment with Atenolol will often reduce the severity of the left ventricular outflow tract obstruction. If needed, treatment for heart failure with Lasix and Enalapril can be used.

Tricuspid Valve Dysplasia

Tricuspid Valve Dysplasia (TVD) is an uncommon congenital heart defect. It is seen most often in Labrador retrievers, although other breeds of dogs and cats can be affected as well. With TVD, malformation of the tricuspid valve and supporting structures leads to tricuspid regurgitation. This causes secondary right heart dilation and can lead to right sided congestive heart failure. In some cases of TVD, there is also tricuspid stenosis which will contribute to right atrial enlargement and the development of clinical signs. With severe right heart enlargement, arrhythmias such as atrial fibrillation can develop. Most dogs with TVD are asymptomatic at diagnosis. However, the murmur is not always detected at a young age due to variable murmur intensity and a PMI on the right. Patients with severe TVD typically will develop right sided heart failure. However the age of onset is quite variable, ranging from 1-7 years. Once heart failure develops, the patients can typically be maintained with an acceptable quality of life using a combination of medications and intermittent abdominocentesis for 1-2 years.