This issue's specialist column on congenital cardiac disease is broken into two parts. This instalment discusses surgical interventions for some of the more common conditions, and the second, which will appear in the spring 2021 issue, will focus on pacemakers.

INTERVENTIONS FOR CONGENITAL CARDIAC DISEASE

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Bea, a 16-week-old female Landseer Newfoundland, was presented to a veterinarian as part of a litter examination before being sold. Out of six puppies, murmurs were detected in three, including Bea. Murmurs were described as being at the left heart base, grade 3 or higher, suggesting that they were indicative of congenital cardiac disease. Bea's veterinarian performed thoracic radiographs, revealing marked cardiac enlargement and possible early left-sided congestive heart failure. The three affected puppies were promptly referred to a cardiologist, at which time Bea was diagnosed with a patent ductus arteriosus (PDA) and severe subaortic stenosis (SAS). One of the other puppies had mild pulmonary valve stenosis, which was not felt to be an issue that would affect the dog's quality of life or longevity; however, the third puppy was diagnosed with a PDA and severe pulmonic stenosis. Bea and her other severely affected littermate had guarded long-term prognoses, and the breeders indicated that they might consider euthanasia for these puppies.

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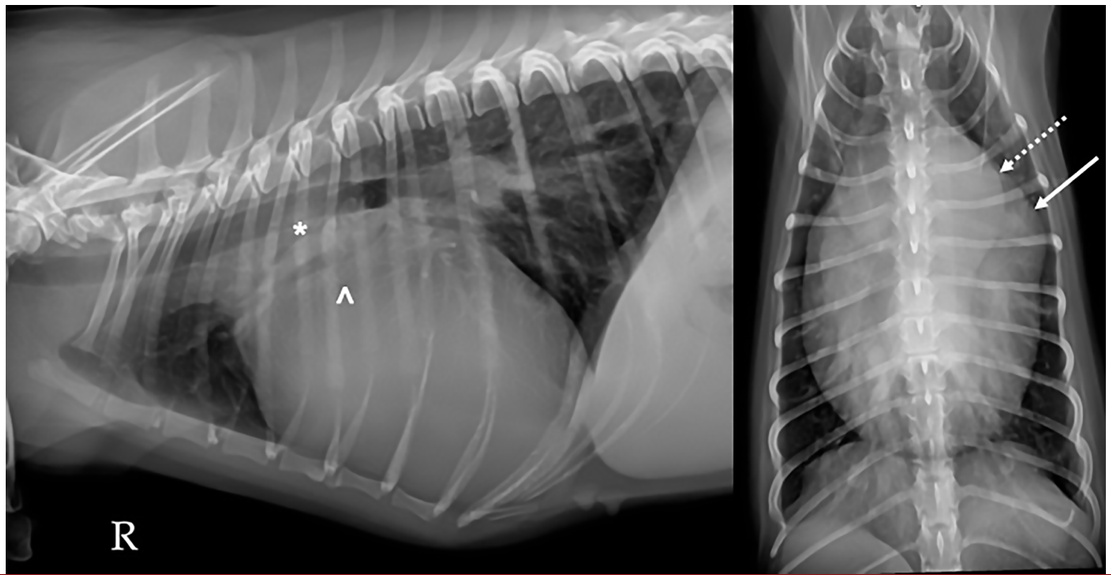

FIGURE 1: Right lateral and VD thoracic radiographs from a dog with a PDA. The cardiac silhouette is markedly enlarged with predominantly left-sided enlargement. On the VD projection, there is dilation of the pulmonary artery (dashed arrow) and left auricle (solid arrow). The pulmonary arteries (*) and veins (^) are both dilated. There is a hypervascular lung pattern, but no evidence of congestive heart failure.

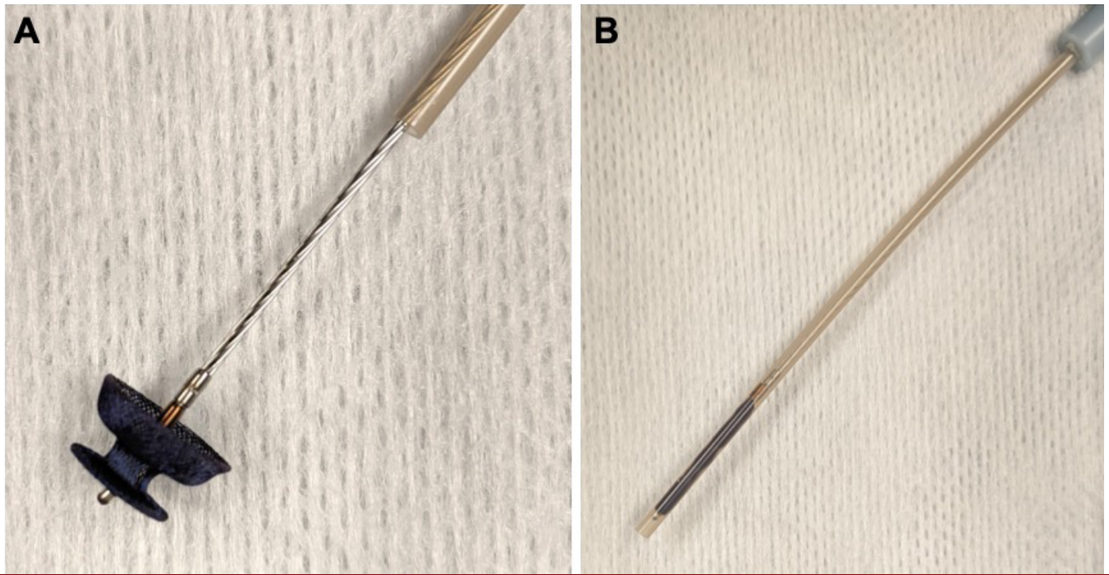

FIGURE 2: An Amplatz canine ductal occluder (ACDO). The ACDO is a nitinol mesh that consists of a flat distal disc that sits in the pulmonary artery and a proximal cup that sits in the ductus (A). The device can be constrained into a catheter for minimally invasive deployment (B).

My wife was a veterinary student in the cardiology rotation when the puppies came in. Knowing that Bea might have a guarded prognosis and might only live a couple of years, my wife told the breeders that we would take Bea, who we renamed Juneau. The cardiologist repaired Juneau's PDA with a minimally invasive procedure that was relatively novel at the time. Correcting her PDA had the effect of also markedly reducing the volume in her left ventricle, which reduced her SAS severity from severe to mild. Her heart failure also immediately resolved.

Juneau lived with us for 10 years, through the remainder of veterinary school, internships, residencies, and early life as specialists. And she is one of the major reasons that I decided to become a cardiologist. The quick action of the initial veterinarian who examined her was crucial in ensuring that she had such a long, healthy life.

Congenital cardiac disease in dogs was historically treated with invasive thoracotomies and open-heart surgeries that entailed major risks. These are still needed in select situations, but there have been major advancements over the past 10 to 15 years in minimally invasive catheter-based procedures to address these issues, called interventional cardiology. In this article, I will discuss two of the most common congenital abnormalities in dogs and the current interventional approaches we take for these conditions.

PDAs

The ductus arteriosus (DA) is a normal fetal connection between the aorta and pulmonary artery that allows blood to bypass the non-inflated lungs. At birth, lung expansion leads to a rapid drop in the resistance to pulmonary blood flow and catabolism of placental prostaglandins. The combination of these events leads to vasoconstriction of the smooth muscle within the DA. Functional closure of the DA generally occurs within hours of birth, with complete closure within a few days.

In dogs, a lack of smooth muscle within the ductus arteriosus can lead to abnormal patency into post-natal life, called a patent ductus arteriosus (PDA). Since systemic pressures (approximately 120/80 mmHg) are much higher than pulmonary pressures (approximately 25/10 mmHg), this results in shunting of blood from the high-pressure aorta to the low-pressure pulmonary artery (i.e., left to right). The shunted blood is sent through the lungs and back to the left heart chambers. This can result in left-sided volume overload.

PDAs are one of the three most common congenital cardiac defects in dogs, accounting for roughly 30 per cent of congenital cardiac diseases. Females are roughly three times as likely to have PDAs as males. Breed predispositions include: Miniature Poodles, Maltese, Bichon Frise, Newfoundlands, and German Shepherds, although any breed can have a PDA. PDA is a hereditary condition; as such, affected dogs should not be bred.

Auscultation high in the left axilla reveals a continuous murmur because the pressure in the aorta is higher than the pulmonary artery in both systole and diastole. The persistent runoff of blood across the ductus during diastole leads to a drop in the diastolic systemic blood pressure, leading to hyperkinetic or "bounding" femoral pulses. Most dogs are asymptomatic at the time of diagnosis; however, if a PDA is detected later, signs of congestive heart failure may be present. Early detection before development of clinical signs is key to ensuring the best prognosis for these patients. By the time a puppy is presented for its first wellness exam, a continuous murmur should not be present. If present, this should prompt referral for cardiac evaluation.

The left-to-right shunt creates a volume overload of the left heart chambers. Over time, this can lead to development of congestive heart failure. Thoracic radiographs can show marked cardiomegaly predominated by left-sided enlargement (Figure 1). On the VD projection, a classic "triple bubble" appearance has been described, with three dilations in the one, two, and three o'clock regions that correspond to aortic enlargement, pulmonary artery enlargement, and left auricular enlargement, respectively. Pulmonary arteries and veins may both be dilated due to pulmonary overcirculation. The pulmonary parenchyma can be challenging to interpret due to a hypervascular lung pattern that can look like early pulmonary edema. More advanced edema will cause interstitial-to-alveolar pulmonary infiltrates, often worst in the perihilar region with extension into the caudodorsal lung fields.

Without treatment, approximately 65 per cent of dogs will develop signs of congestive heart failure within the first year of life, with the remaining dogs developing heart failure as juveniles or young adults. It is uncommon for a dog with a PDA to not eventually develop congestive heart failure. Supraventricular (e.g., atrial fibrillation) and ventricular arrhythmias can also result from marked chamber enlargement. Given these risks, correction is recommended almost universally for dogs with left-to-right PDAs. Rarely, dogs may develop severe pulmonary hypertension such that the shunt reverses (i.e., from pulmonary artery to aorta, or right to left), and the murmur disappears. Shunt reversal generally occurs in dogs before six months of age. Surgical correction is not recommended if this occurs, as occlusion of the PDA in this situation generally leads to fulminant right heart failure signs due to the severe pulmonary hypertension.

Historically, dogs with PDAs required a thoracotomy to ligate the vessel; however, over the last several years transcatheter deployment of an occlusion device has become the preferred method in most cases. The most common device used is called an Amplatz canine ductal occluder (ACDO) (Figure 2). The smallest ACDO requires at least a 4 Fr delivery catheter to be fitted into the femoral artery. For this reason, there is a lower weight limit for performing this procedure: generally around 3 kg. Less commonly used occlusion devices such as coils or vascular plugs may be usable in dogs weighing less than 3 kg for minimally invasive closure. A traditional thoracotomy could also be performed in these dogs; however, tearing of the ductus during ligation could be fatal.

For transcatheter occlusion of a PDA, a cut-down is performed in the inguinal region to access the femoral artery. Similar to placement of a central (jugular) venous catheter, a modified Seldinger technique is used to facilitate placement of a long introducer. Under fluoroscopic guidance, the introducer is advanced through the descending aorta until it reaches the juncture between the transverse and descending aorta. Angiography is performed via injection of radiopaque contrast, which not only delineates the position and morphology of the PDA, but also allows confirmative measurements used for device sizing (Figure 3). A guide wire is subsequently fed through the introducer and across the ductus into the main pulmonary artery, and the introducer is then tracked into the same position. The ACDO is then deployed through the introducer to occlude the PDA. A second angiogram is performed through the introducer to confirm occlusion

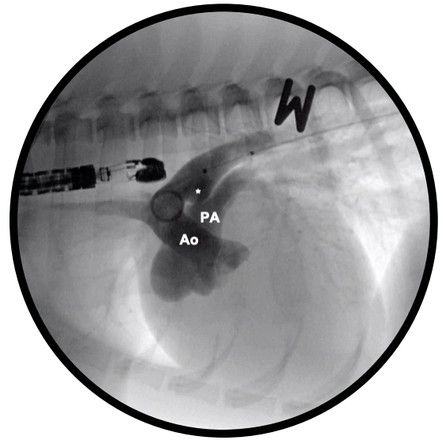

FIGURE 3: Aortic angiogram in a dog with a patent ductus arteriosus. A pigtail angiographic catheter with radiopaque markers (for measurement calibration) has been advanced through the descending aorta with the pigtail sitting in the aortic arch (Ao). Contrast injection reveals a left-to-right shunting patent ductus arteriosus (*) with subsequent opacification of the main pulmonary artery (PA). A transesophageal echocardiographic probe is also visible.

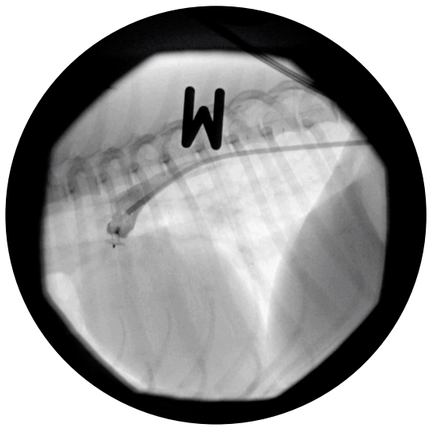

FIGURE 4: Aortic angiogram several minutes after deployment of an ACDO. The contrast extends to the proximal cup of the ACDO but does not cross into the pulmonary artery, indicating complete occlusion of the PDA.

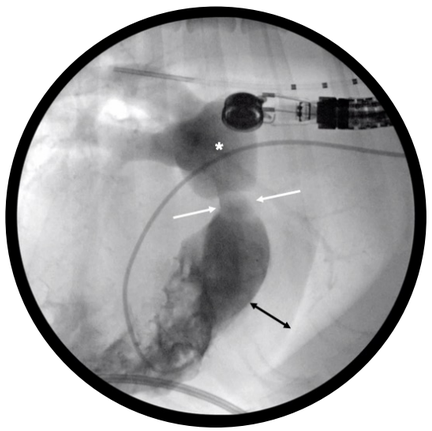

FIGURE 5: Right ventricular angiogram in a dog with pulmonary valve stenosis. Orientation is such that the patient's head is to the right to coincide with the surgeon's approach. A pigtail angiographic catheter has been advanced through the jugular vein, cranial vena cava, and right atrium with the pigtail resting in the right ventricular outflow tract. Contrast injection reveals a discrete narrowing at the level of the pulmonic valve (white arrows) with post-stenotic dilation of the main pulmonary artery and extending into the branch pulmonary arteries (*). Severe right ventricular hypertrophy is also evident (black arrow). A transesophageal echocardiographic probe and marker catheter (for measurement calibration) are also visible.

of the ductus (Figure 4). Once this is confirmed, the ACDO can be unscrewed from the delivery wire. All catheters are then removed, and the femoral artery is ligated before closure.

Complications of transcatheter PDA occlusion are uncommon, but can include bleeding, dislodgement of the device, and infection. If the device dislodges and travels into the peripheral lungs, animals may show no clinical signs aside from recurrence of the heart murmur and potentially heart failure if those signs were present preoperatively. Systemic embolization of the device is a surgical emergency to restore blood flow. Infection can be particularly catastrophic as there is no feasible way to remove the device from the ductus.

Barring complications, prognosis is excellent in dogs undergoing transcatheter PDA occlusion. Dogs with no prior history of congestive heart failure are expected to live a normal life, emphasizing the importance of early referral and intervention in these cases. Dogs with a history of heart failure can often still have a good prognosis if correction can be performed shortly after diagnosis, with many dogs being weaned off cardiac medications.

BALLOON VALVULOPLASTY

Pulmonic stenosis (PS) is also one of the most common congenital cardiac diseases in dogs with a frequency similar to PDAs. A range of lesions have been described with this condition, including leaflet fusion, dysplastic thickened leaflets, and hypoplasia of the pulmonic valve annulus. Bulldog breeds may also have an aberrant left coronary artery that wraps around the pulmonic valve region and causes extramural compression. PS creates a pressure overload in the right ventricle leading to thickening of the right ventricular walls. This can lead to development of arrhythmias and right-sided congestive heart failure. Because the arrhythmias tend to originate in the ventricles, sudden death is possible. Dogs with PS are often asymptomatic as puppies but can go on to develop signs such as activity intolerance or exertional syncope at any point in adulthood.

Early identification of PS is important, but may be challenging. Heart murmurs associated with PS are best heard in the left base region, and softer murmurs can be difficult to distinguish from innocent puppy murmurs. Innocent puppy murmurs are generally grade 3 or lower intensity, systolic, dissipate at each visit, and are usually gone by the age of 16 weeks of age. A murmur with greater than grade 3 intensity, increasing in intensity, in an unusual location, or with a diastolic or continuous component is never normal and always warrants further evaluation. Other physical examination abnormalities in dogs with PS could include arrhythmias or signs of right-sided congestion, such as jugular venous distension, hepatomegaly, and ascites.

Symptomatic patients should have routine lab work and thoracic radiographs performed. Definitive diagnosis is obtained from an echocardiogram, which also helps to quantify the severity of the stenosis. In addition to diagnosis of PS, the goal of the echocardiogram is to identify any other congenital cardiac abnormalities, quantify right-sided enlargement, and obtain precise views of the valve, annulus, and the amount of room available for a balloon to fit. Aberrant coronary arteries in bulldogs can generally not be excluded via the echocardiogram, but can be evaluated with either thoracic CT or an aortic root angiogram at the time of surgery.

Patients with mild PS generally do not require any therapy and are expected to live normal lives. Patients with severe PS are at the highest risk for developing complications of the disease, and generally require therapy. Patients with moderate PS have mixed outcomes, and treatment is based upon the presence of clinical signs, severity of the disease, presence of other cardiac abnormalities, and morphologic valve changes that may help predict procedural success. PS dogs with concurrent tricuspid valve dysplasia generally require treatment. Dogs that have a combination of PS and a ventricular septal defect (VSD) may not be treated if the PS is decreasing the shunting across the VSD.

Beta blockers (e.g., atenolol) are often prescribed for moderate and severe PS cases. By decreasing the force of contraction and heart rate, they help to lessen the degree of obstruction and decrease the myocardial demand for oxygen. They are also antiarrhythmic for both supraventricular and ventricular tachyarrhythmias. I usually aim for a target dose of 1 mg/kg PO BID for atenolol, but will slowly up-titrate to this dose over one to three weeks.

Balloon valvuloplasty has been shown to improve clinical signs and improve survival in dogs with PS, and has therefore become the preferred surgical treatment. This is a minimally invasive procedure performed from either the jugular vein or the femoral

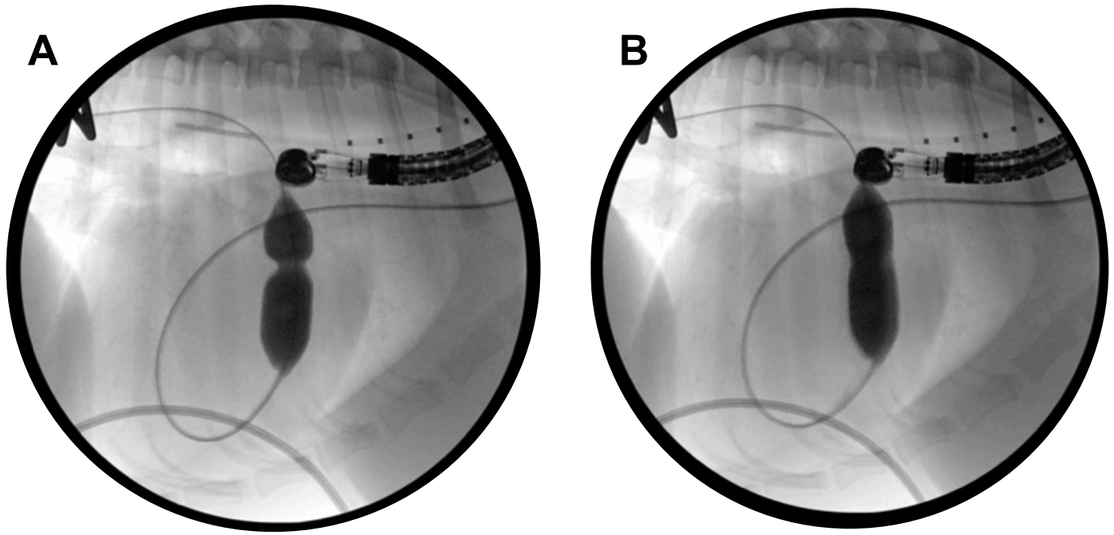

FIGURE 6: Serial images during balloon inflation for pulmonary valve stenosis. In both images, the patient's head is to the right so as to be oriented with the surgeon's approach. A guide wire has been fed from the jugular vein through the right heart and distally in the pulmonary artery so as to allow placement of a balloon catheter in the right ventricular outflow tract. A: Initial inflation reveals a discrete waist at the site of pulmonic valve stenosis. B: Continued inflation results in an abrupt loss of the balloon waist as the pulmonic valve leaflets tear open.

vein, sometimes being performed percutaneously without the need for any incision. First, a vascular introducer is placed into the vein. An angiographic catheter is advanced into the right ventricle under fluoroscopic guidance, and contrast injection helps to delineate the stenosis and confirm annulus measurements obtained from the echocardiogram (Figure 5). If an aberrant coronary artery is identified on the angiogram, the procedure is either aborted or a more conservative ballooning is performed. A guide wire is then fed into the distal pulmonary artery and used to facilitate placement of an inflatable balloon in the right ventricular outflow tract. On initial inflation, a discrete waist will initially be seen and with continued inflation, the goal is to have this waist abruptly disappear, indicating tearing of the valve leaflets (Figure 6). On subsequent inflations, a waist will not be present if this has occurred.

Complications of balloon valvuloplasty include damage of other cardiac structures such as the tricuspid valve or perforating a heart chamber. Ventricular arrhythmias can be very frequent during the procedure and lead to ventricular fibrillation if not treated or not responding to treatment intraoperatively. These almost invariably subside once catheters are removed from the heart. With inflation of the balloon, it is not uncommon to damage the right bundle branch, creating a right bundle branch block (Figure 7). This may be transient or even heart rate–dependent, although it is not generally of any significant concern. If permanent, it is important to know that future ECGs performed for anesthesis may look unusual.

The general goal of balloon valvuloplasty is to reduce the pressure gradient by as much as safely possible while minimizing damage to any other cardiac structures. Published definitions of procedure success include a 50 per cent reduction in PS severity, or reduction out of the severe category of PS. Procedural success rates are 80 per cent to 90 per cent in the literature, but it is important that the procedure be performed before development of major complications to ensure the best prognosis. Arrhythmias present before surgery may be irreversible, and although reduction may help the animal in right-sided congestive heart failure, some degree of medical management may be required beyond surgery. WCV

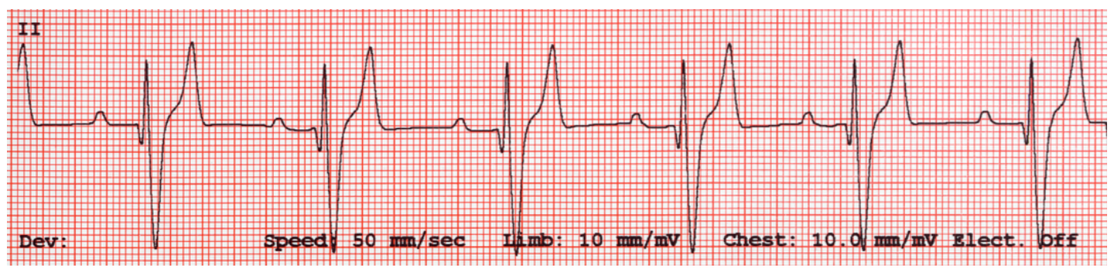

FIGURE 7: Lead II ECG from a dog post–balloon valvuloplasty. The P waves are normal in appearance with a set relationship to the subsequent QRS complexes, indicating a sinus rhythm. The QRS complex is widened and predominantly negative due to the presence of a deep S wave. These changes indicate ventricular conduction is occurring with a right bundle branch block, a common finding after balloon valvuloplasty that may be transient or permanent.