Current management of canine congenital heart disease
ETVMA: 09/26/2021

Jonathan A. Abbott, DVM, Dipl. ACVIM (Cardiology)
University of Tennessee,
Knoxville, TN

Introduction

This session is intended to provide a synopsis of the current diagnostic and therapeutic approach to congenital heart disease (CHD). The management of *canine* CHD, including the use of minimally invasive, transcatheter techniques in the most commonly occurring malformations will be emphasized.

Etiology of CHD

The cause of canine CHD is largely unknown although a genetic basis has been proven for a few specific malformations. Barring identification of a genetic mutation that is consistently associated with an abnormal phenotype, planned breeding studies or careful evaluation of accurate pedigrees are necessary to demonstrate genetic transmission of a congenital malformation. Indeed this has been accomplished for a few defects. For example, subvalvular aortic stenosis is an inherited trait in Newfoundland dogs, genetic transmission of pulmonic stenosis has been demonstrated in the beagle hound and a spectrum of conotruncal malformations that includes Tetralogy of Fallot is inherited in keeshonden. Pronounced breed predispositions are recognized for some forms of canine CHD and in these cases, it is probable that the defect has a genetic basis.

Recognition and Diagnosis of Congenital Heart Disease

The patient history of those with CHD rarely provides specific findings and the majority of canine patients with CHD are free of clinical signs when the disorder is first detected. Importantly, normal growth and lack of clinical signs do not imply that CHD is of no clinical importance nor necessarily imply a favorable long-term prognosis. This serves to emphasize the importance of accurate diagnosis even for cases in which clinical signs are absent.
The vast majority of cardiac malformations result in cardiac murmurs. Therefore, congenital heart disease usually is first identified when outwardly healthy puppies or kittens are presented for routine veterinary evaluation. It is noteworthy that some normal puppies and kittens have murmurs that do not result from cardiac disease. These murmurs, known as innocent murmurs, are soft, always systolic, and usually heard best over the left heart base. The intensity of an innocent murmur may vary from day to day or even from moment to moment in association with changes in heart rate. Murmurs that are innocent generally become inaudible before the patient is 8 months of age. While congenital disease can result in a soft murmur, a loud cardiac murmur, or one that is diastolic or continuous, invariably suggests the presence of cardiac disease and further diagnostic investigation is indicated. Further evaluation may include electrocardiography and thoracic radiography but these tests rarely provide diagnostically specific information. Doppler echocardiography performed by an experienced examiner provides a definitive, non-invasive diagnosis in practically all cases of CHD.

THERAPY OF CONGENITAL HEART DISEASE

Optimally, treatment of CHD is through surgical methods or interventional cardiac catheterization techniques. Medical therapy is apt to be palliative only.

Surgical Techniques / Cardiopulmonary Bypass

The availability of cardiopulmonary bypass - or rather, its lack - is one factor that limits the effective management of CHD in veterinary medicine. A few defects can be surgically managed without the need for bypass; a PDA for example, can be ligated without entering the circulation. However, defects that require access to the left ventricle and/or prolonged manipulation can only be performed with cardiopulmonary bypass. Surgical procedures that require cardiopulmonary bypass currently are performed only at a few veterinary institutions in North America and elsewhere.

Interventional Catheterization Techniques

Originally, cardiac catheterization - the art and science of manipulating catheters within the cardiovascular system - was a diagnostic technique. Usually under fluoroscopic guidance, catheters can be used to selectively deliver dye, to measure blood oxygen contents and to directly measure intracardiac pressures. Beginning in the 1960's, a number of resourceful pediatric cardiologists introduced catheterization techniques that were intended to treat or "intervene". The techniques that are used most often in veterinary cardiology are pulmonic balloon valvuloplasty and transcatheter occlusion of patent ductus arteriosus

PATENT DUCTUS ARTERIOSUS (PDA)

The ductus arteriosus (DA) connects the ventral aspect of the descending aorta to the bifurcation of the main pulmonary artery. During fetal life, the DA diverts the majority of the right ventricular stroke volume to the aorta. In normal individuals, closure of the ductus occurs within days of birth; the process is complex but involves a prostaglandin cascade. Failure of the duct to close, which in most cases is explained by a lack, or relative lack, of ductus specific smooth muscle, results in a persistently patent DA or, PDA. A genetic basis for failed closure of the DA has been documented in miniature / toy poodles. When the duct is the only defect, and pulmonary vascular resistance decreases following birth, blood shunts from the high pressure / high resistance systemic circulation to the low pressure / low resistance pulmonary circulation. Therefore, the shunt direction is from left-to-right; this increases pulmonary blood flow *and* pulmonary venous return imposing a volume load on the left atrium and ventricle. The development of myocardial dysfunction, mitral valve regurgitation and pulmonary edema are potential consequences of the shunt. PDA results in a continuous murmur; that is, a murmur that begins in systole and continues**,** without interruption**,** into diastole. When the shunt is substantial, the arterial pulse is hyperkinetic or, "bounding". Electrocardiography often discloses evidence of left ventricular hypertrophy. Radiographically, there is left-sided cardiomegaly that is roughly commensurate with the size of the shunt**.** Additional findings may include prominence of the main pulmonary artery, proximal aorta and left atrium. The diagnosis is confirmed echocardiographically. Specific findings of course depend on the size of the shunt but typically include left atrial and left ventricular enlargement. Doppler examination provides evidence of a continuous flow disturbance within the main pulmonary artery. The ductus, and certainly the ductal orifice of the pulmonary artery**,** can be identified in almost all cases.

Therapy

A minority of patients have a small, well-tolerated duct but usually, intervention is indicated when a PDA is identified in a dog that is younger than 24 months old. Surgical ligation can be performed without cardiopulmonary bypass and though minimally invasive transcatheter occlusion has become routine, surgical ligation remains an appropriate therapeutic approach that is associated with low mortality.

Transcatheter PDA occlusion using different devices and subtly different techniques has been reported. Thrombotic Gianturco coils were widely used until two veterinary cardiologists, Ngyuenba and Tobias, in collaboration with a manufacturer of cardiovascular devices, developed a metallic plug, the ACDO, or Amplatz Canine Ductal Occluder, that was specifically designed to occlude the canine ductus. Use of this device has almost completely supplanted the use of the Gianturco coil in veterinary practice. The ACDO can be used to successfully occlude PDA over the broad range of ductal size and morphologies.

Numerous variations on the basic technique of transcatheter ductal occlusion have been reported. Most often the devices are deployed within the ductus after retrograde catheterization of the aorta. Briefly, after induction of general anesthesia, access to the femoral artery is most often obtained after a small inguinal incision but vascular access can be percutaneously obtained using the modified Seldinger technique. Using fluoroscopic guidance, an angiographic catheter and/or vascular sheath is advanced to the ascending aorta and an angiogram recorded after injection of contrast material in the proximal descending aorta. Then, the device is advanced through a catheter or vascular delivery sheath and deployed within the ductus. Major complications of transcatheter intervention for PDA include intra-operative death, incomplete occlusion, post-procedural hemolysis, and coil migration. Mortality associated with transcatheter intervention for PDA generally is quite low, near 2%, although higher mortality has been reported in small studies that specifically recruited high risk patients. Body size is an important determinant of the suitability of the technique. Because of the size of the delivery devices, transcatheter occlusion of PDA in patients that weight less than 3 kg is problematic.

SUBVAVULAR AORTIC STENOSIS (SAS)

In dogs, left ventricular outflow tract obstruction most commonly results from the presence of a subvavular fibrous or fibrocartilaginous ring that develops in the first weeks of life. The pressure gradient across the obstruction, which can be measured - by cardiac catheterization - or estimated - by Doppler echocardiography - is used as a clinical measure of stenosis severity. Pressure gradients that are less than 40 mmHG are mild and those greater than 100 mmHg are severe; intermediate gradients are described as moderate. Aortic stenosis is most common in large breed dogs including Golden retrievers, Rottweilers, as well as Boxer, Newfoundland and German Shepherd dogs. Clinical signs in puppies are uncommon; syncope and sudden death are observed in young adults with severe obstructions.

Cardiopulmonary bypass is required for surgical repair. Although a new technique in which a “cutting balloon” is used might have promise, published data suggest that neither surgical correction nor balloon dilation improve survival relative to medical therapy consisting atenolol. Evidence that administration of atenolol is superior to placebo or no treatment is lacking.

PULMONIC STENOSIS (PS)

Right ventricular outflow tract obstruction usually results from valvular dysplasia. PS occurs commonly in terriers, English Bulldogs, miniature schnauzers and Samoyeds.

Therapy

PS can be treated surgically but is more often addressed by transcatheter balloon dilation. The pressure gradient that represents an indication for intervention is not known with precision although it is known that balloon dilation confers a survival benefit for those with gradients that exceed 80 mmHg. Balloon dilation of PS is performed under general anesthesia. After aseptic preparation of the groin or cervical region, access to the jugular or femoral vein is typically obtained percutaneously. After hemodynamic and angiographic studies, an end-hole diagnostic catheter is guided fluoroscopically into the pulmonary artery. The end-hole catheter is then exchanged for a balloon dilation catheter over a long wire-guide. Balloon catheters are available from manufacturers in numerous sizes; appropriate dimensions are determined by the size of the patient and echocardiographic (or angiographic) measurement of the pulmonic valve annulus. Several inflations with saline-diluted contrast material are performed after the balloon has been positioned across the stenosis. The inflation is observed fluoroscopically; the obstructive valve results in the appearance of a “waist”; ideally, the waist disappears suddenly during the first inflation and is not observed during subsequent attempts. After balloon inflation, catheters are withdrawn and patient is recovered. Most patients are discharged the day after the procedure which permits echocardiographic re-evaluation after complete recovery from anesthesia. Success seems to depend to a great extent on the nature of the stenosis; patients with isolated valvular stenosis in which there is fusion of otherwise normal valve cusps tend to benefit the most from the procedure.

VENTRICULAR SEPTAL DEFECT

A defect of the interventricular septum results in a communication between the left and right ventricles. Presumably because of its complex embryonic derivation, most ventricular septal defects (VSD) involve the membranous part of the septum; generally these defects are subaortic with a right ventricular orifice that is immediately subjacent to the septal tricuspid leaflet. The clinical importance of the defect depends on: the size of the defect and, the presence or absence of other concurrent defects. When the VSD is the only cardiovascular lesion, blood shunts from left-to-right; this increases pulmonary blood flow and pulmonary venous return resulting in a volume load on the left atrium and left ventricle. A VSD results in a systolic murmur; this is because the pressure difference between the ventricles drops to nearly zero during diastole.

Therapy

Definitive repair requires cardiopulmonary bypass but pulmonary artery banding is occasionally used as a palliative surgical intervention. Transcatheter occlusion of VSD is routinely practiced in pediatric cardiology and transcatheter closure in dogs has been reported. Most VSD in dogs are small, well tolerated and do not require treatment.

Selected References – complete references available from author

Gordon SG, Miller MW. Transarterial Coil Embolization for Canine Patent Ductus Arteriosus Occlusion. Clinical Techniques in Small Animal Practice 2005;20:196-202.

Johnson MS, Martin M, Edwards D, et al. Pulmonic stenosis in dogs: balloon dilation improves clinical outcome. J Vet Intern Med 2004;18:656-662.

Locatelli C, Spalla I, Domenech O, et al. Pulmonic stenosis in dogs: survival and risk factors in a retrospective cohort of patients. Journal of Small Animal Practice 2013;54:445-452.

Meurs KM, Lehmkuhl LB, Bonagura JD. Survival times in dogs with severe subvalvular aortic stenosis treated with balloon valvuloplasty or atenolol. Journal of the American Veterinary Medical Association 2005;227:420-424.

Nguyenba TP, Tobias AH. The Amplatz(R) canine duct occluder: A novel device for patent ductus arteriosus occlusion. Journal of Veterinary Cardiology 2007;9:109-117.

Nguyenba TP, Tobias AH. Minimally Invasive Per-Catheter Patent Ductus Arteriosus Occlusion in Dogs Using a Prototype Duct Occluder. Journal of Veterinary Internal Medicine 2008;22:129-134.

Orton EC, Herndon GD, Boon JA, et al. Influence of open surgical correction on intermediate-term outcome in dogs with subvalvular aortic stenosis: 44 cases (1991-1998). Journal of the American Veterinary Medical Association 2000;216:364-367.

Singh MK, Kittleson MD, Kass PH, et al. Occlusion Devices and Approaches in Canine Patent Ductus Arteriosus: Comparison of Outcomes. Journal of Veterinary Internal Medicine 2012;26:85-92.