Congenital heart disease (CHD) is defined as a morphologic defect of the heart or associated great vessels present at birth. Abnormalities are caused by alterations or arrests in particular phases of embryonic development of the fetal heart. The term *congenital* does not imply that the defect was inherited, and the defect may have occurred spontaneously or secondary to a drug or toxin. By studying families of animals with specific CHDs, many defects have also been shown to be heritable. Additionally, if the defect was caused by a spontaneous de novo mutation, that individual has the potential to transmit the mutation to offspring. The diagnosis of CHD is important not only to the health of the patient but to eliminate affected individuals from the breeding pool.

**INCIDENCE**

Using the University of California, Davis, Veterinary Medical Teaching Hospital database, approximately 17% of dogs and 5% of cats examined by the cardiology service over a 10-year period were diagnosed with CHD [1]. The most common CHDs were subaortic stenosis (SAS) and patent ductus arteriosus (PDA) in dogs and tricuspid valve dysplasia (TVD) and ventricular septal defect (VSD) in cats.

**CLINICAL APPROACH**

The clinical approach to diagnosis of CHD follows the same fundamental principles as with acquired cardiac diseases. Signalment is important in the evaluation of CHD, because many defects have particular breed predispositions (Table 1). A history of familial cardiac disease is also important to obtain to establish a possible heritable basis of CHD. Most young animals with CHD are asymptomatic when first examined, and presence of a heart murmur is often the first clue of CHD. Syncope or exercise intolerance may be seen, and they may be ominous signs of significant cardiac disease. Cyanosis may be
identified in animals with pulmonary-to-systemic shunting defects (ie, right-to-left shunts). These animals may also have stunted growth compared with the rest of the litter. Dyspnea, tachypnea, or cough may be identified as a result of congestive heart failure.

PHYSICAL EXAMINATION
The first step to identifying a puppy or kitten with CHD is detection, localization, and characterization of a murmur (Table 2). Often, the location of a murmur and its timing may be pathognomonic for a defect or may refine the differential list of possible defects. Detection of a left basilar continuous murmur is pathognomonic for a PDA. A murmur is present in most animals with CHD, with a few exceptions, including a right-to-left shunting PDA or Eisenmenger’s syndrome secondary to a large left-to-right shunt. Soft systolic murmurs (grade I–II/VI) may be heard with innocent physiologic flow murmurs because of increased flow velocity in the aorta or pulmonary artery in pediatric patients, but such murmurs usually disappear by 6 months of age. Innocent murmurs may be dynamic and vary in intensity, based on the level of activity or excitement. The murmur intensity in certain defects, such as SAS, may worsen over the first several months of life.

Other important physical examination abnormalities are alterations in femoral arterial pulses; cyanosis (generalized versus differential); signs of right heart
failure (ie, jugular venous distention, ascites, or hepatomegaly) and respiratory abnormalities (ie, dyspnea or increased adventitious lung sounds). Hyperkinetic bounding pulses (ie, waterhammer) are felt when there is significant aortic diastolic runoff with a PDA or severe aortic insufficiency. Conversely, dampened hypokinetic arterial pulses may be felt when there is obstruction to blood flow out of the aorta secondary to moderate to severe SAS or when there is markedly diminished cardiac output. Cyanosis occurs when the PaO₂ is 45 mm Hg or less, and this should raise the clinical suspicion of pulmonary-to-systemic shunting defects.

**LABORATORY TESTS**
A complete blood cell count (CBC) and chemistries are often unremarkable in animals with CHD. Animals with pulmonary-to-systemic shunting defects often have polycythemia and/or hypoxemia and may have metabolic acidosis.

**THORACIC RADIOGRAPHS**
Evaluation of cardiac size, great vessels, and pulmonary vasculature is useful to identify patients with moderate or severe CHD. Combined with physical
examination abnormalities, such as a murmur and femoral pulse characteristics, radiographs may be useful to narrow the differential diagnosis list. Radiographs are often normal if there is only mild CHD. Cardiac size in patients with SAS is usually normal, and thoracic radiographs may be unremarkable even in the presence of severe disease. Examination of the great vessels often reveals poststenotic vasodilation of the aorta or pulmonary artery in patients with aortic stenosis or pulmonic stenosis (PS), respectively (Figs. 1 and 2). The presence of an aortic bulge (ductal bump) near the origin of the ductus is pathognomonic for a PDA (Fig. 3). Pulmonary vascular overcirculation (ie, distention of pulmonary arteries and veins) may be detected in patients with significant systemic-to-pulmonary shunts, including a PDA, VSD, or atrial septal defect (ASD). Diminutive or undercirculated pulmonary vasculature may be seen with pulmonary-to-systemic shunts, such as tetralogy of Fallot (TOF). Animals with moderate or severe TVD often have profound right atrial enlargement and distention of the caudal vena cava (Fig. 4).

**ELECTROCARDIOGRAPHY**

Evidence of a normal electrocardiogram does not rule out CHD. Certain defects, such as PS or TOF, typically result in deep S waves and a right axis deviation. PDA often results in tall QRS complexes indicative of left ventricular hypertrophy. Dogs with severe SAS may have ventricular arrhythmias or ST segment abnormalities indicative of regional myocardial hypoxia. In patients with CHD causing severe atrial enlargement, there may be a supraventricular tachyarrhythmia, such as atrial fibrillation or supraventricular tachycardia.

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**Fig. 1.** Thoracic radiographs of a dog with SAS. Severe poststenotic dilation of the ascending aorta is seen on the lateral (A) and dorsoventral (B) radiographs in a dog with SAS. The heart size is normal, because there is concentric hypertrophy of the left ventricle.
ECHOCARDIOGRAPHY

Echocardiography is the cornerstone for establishing a definitive diagnosis of a specific CHD. Two-dimensional views are useful to evaluate for atrial dilation, concentric ventricular hypertrophy (ie, in response to pressure overload), or eccentric ventricular hypertrophy (ie, in response to volume overload). Careful evaluation of valvular morphology is essential for diagnosis of
atrioventricular valve dysplasia and valvular stenosis. Color-flow Doppler echocardiography is used to identify and localize the anatomic region of turbulent blood flow and may be followed by pulsed-wave and continuous-wave Doppler echocardiographic measurement of blood flow velocity. Noninvasive estimation of the pressure gradient between two regions is possible by measuring maximal blood flow velocity (using continuous-wave Doppler echocardiography) and is converted into pressure units by the modified Bernoulli equation:

\[
\text{pressure gradient between two chambers} = 4 \times \text{velocity (m/s)}^2
\]

Fig. 3. Thoracic radiograph and aortic angiograms of a dog with a PDA. (A) Pathognomonic radiographic sign of a PDA is the ductal aneurysm (ie, ductal bulge) of the aorta on the dorsoventral radiograph. There is severe left-sided cardiomegaly and mild pulmonary overcirculation. (B) Aortic root angiogram shows the blood shunting from the aorta (Ao), through the ductus (D), and into the pulmonary artery (PA). (C) After a transarterial thrombogenic coil was placed in the ductus, another angiogram is performed, which shows a coil properly positioned within the ductus, resulting in complete ductal closure with no dye passing into the pulmonary artery.
The estimation of pressure gradients is essential for determination of the severity of obstructive lesions or evaluation of pulmonary hypertension secondary to several CHDs. Positive-contrast echocardiography using agitated saline injected into a peripheral vein is a sensitive method to evaluate for the presence of right-to-left intracardiac or extracardiac shunting defects. Normally, microbubbles enter the right atrium and pass through the right ventricle and pulmonary artery to be filtered by the pulmonary capillaries. The presence of microbubbles within the left heart or aorta confirms a right-to-left shunt, and determination of the region of the right-to-left shunt is possible. For example, if microbubbles appear in the right atrium and then are seen within the left atrium, there is a right-to-left shunting ASD or patent foramen ovale.

**CARDIAC CATHETERIZATION AND ANGIOGRAPHY**

Given the advances in echocardiography, cardiac catheterization is rarely necessary to achieve a clinical diagnosis. Often, cardiac catheterization with pressure measurements and angiography are performed before interventional techniques for the treatment of specific CHDs, such as PS balloon valvuloplasty or percutaneous PDA arterial coil embolization. Measurement of cardiac output as well as peripheral and systemic vascular resistance and determination of the ratio of pulmonic-to-systemic blood flow (QP/QS) may be helpful to identify good candidates for surgical treatment for several left-to-right shunting CHDs (QP/QS >2.5). Surgical treatment is indicated if the main cause of the pulmonary hypertension is increased pulmonary blood flow rather than
increased pulmonary vascular resistance. Right heart catheterizations are most commonly performed via a jugular venotomy or may be performed from the femoral vein. Left heart catheterizations are performed via the carotid artery or the femoral artery, depending on which interventional procedure is planned (ie, the right carotid artery is used mostly for balloon dilation of SAS, whereas the right femoral artery is most often used for percutaneous coil embolization for treatment of a PDA). Normal peak left ventricular systolic pressure and aortic systolic pressure (~100–120 mm Hg) are four times those of right ventricular pressure and pulmonary arterial systolic pressure (~20–30 mm Hg). Cardiac catheterization is useful to detect an intracardiac pressure gradient in an obstructive lesion, which correlates with the severity of the stenosis.

CLASSIFICATION OF CONGENITAL HEART DISEASES

Valvular Obstructive Diseases

Subaortic stenosis

SAS is the most common CHD of large-breed dogs (see Table 1). It is defined as an obstruction of the left ventricular outflow tract caused by a fibrous ridge or fibrotic ring just below the aortic cusps. The defect begins with small fibrous nodules of the endocardium that develop in the first few weeks to months of life, which are likely only possible to detect by pathologic examination. These early lesions are called grade 1 lesions and are rarely accompanied by a heart murmur. Over the first 6 months of life, this ridge grows and causes progressive stenosis of the left ventricular outflow tract. A grade 2 lesion is defined as a narrow fibrotic ridge that extends partially around the left ventricular outflow tract and is often accompanied by a soft grade I to II/VI left basilar systolic murmur. A grade 3 lesion is the most severe and consists of a fibrous band, ridge, fibromuscular tunnel, or fibrotic collar completely encircling the left ventricular outflow tract. There are often focal regions of myocardial necrosis and fibrosis of the subendocardium and inner half of the left ventricle secondary to the pressure overload and ischemia. There is also intramyocardial coronary arteriosclerosis, luminal narrowing, and intimal and medial smooth muscle hypertrophy in the regions of the myocardial necrosis and fibrosis [2].

SAS has been most extensively studied in a breeding colony of Newfoundlands, in which the mode of heritability was most likely autosomal dominant because of a single major gene abnormality [3]. The fibrotic defect is derived from persistent embryonic endocardial tissue within the conotruncal septum, which retains the ability to proliferate and undergo chondrogenic differentiation [2]. Mitral valve dysplasia (MVD) is the most frequent concurrent CHD.

Increased resistance to systolic ejection of blood flow through a narrowed region of the left ventricular outflow tract results in high left ventricular systolic pressure. Concentric left ventricular hypertrophy (ie, thickened walls) occurs to reduce wall stress and compensate for the pressure overload. Systolic function is usually maintained, but there may be diastolic dysfunction when there is chronic, severe, concentric hypertrophy and myocardial fibrosis. Systolic failure may be seen in aged animals with moderate to severe SAS. Myocardial
ischemia may result from reduced density of capillaries within the hypertrophied myocardium, reduced left circumflex coronary artery blood flow, and systolic flow reversal of the intramyocardial coronary blood flow. The subendocardium and papillary muscles are most vulnerable to ischemia.

Exertional syncope and sudden death are common in dogs with severe SAS. Possible mechanisms of exertional syncope include activation of left ventricular pressure receptors resulting in a vagal maneuver (ie, bradycardia ± vasodilation), peripheral arterial vasodilation in the face of fixed resistance to cardiac output, or malignant ventricular tachyarrhythmia secondary to ischemia or sympathetic activation. Malignant ventricular tachyarrhythmias may occur secondary to myocardial ischemia and fibrosis and may lead to sudden death.

Dogs with SAS are likely to die suddenly within the first 3 years of life [4]. Dogs with mild SAS most often have a good prognosis with a normal life expectancy. Older dogs with less severe obstructions may develop CHF failure or aortic valve infective endocarditis. The endothelial surface of the aortic valve is disrupted by the trauma of high-velocity blood flow through the stenotic region, which predisposes dogs to develop infective endocarditis during bacteremic conditions.

The characteristic murmur of SAS is a left basilar holosystolic murmur, with an intensity that roughly correlates with the severity of obstruction. The murmur intensity may worsen over the first few months of life as the fibrotic obstruction worsens. In dogs with severe SAS, the murmur often radiates to the right thorax and carotid arteries. Femoral pulses are often hypokinetic in dogs with moderate to severe SAS because of the delay in peak left ventricular ejection. An electrocardiogram may reveal ST segment elevation or depression (>0.2 mV) indicative of regional myocardial ischemia in dogs with severe SAS. Although there is a wide overlap, ventricular arrhythmias are more common in dogs with severe obstructions. Thoracic radiographs may reveal a poststenotic dilation of the ascending aorta (see Fig. 1). Heart size is usually normal, because the hypertrophy is concentric.

Echocardiography is essential for the diagnosis of SAS and determining the severity of obstruction. Concentric left ventricular hypertrophy and a hyperechoic subendocardium may be seen on two-dimensional echocardiography in dogs with moderate to severe SAS (Fig. 5). The degree of concentric hypertrophy is poorly predictive of the degree of obstruction, however. The ratio of the cross-sectional area of the left ventricular outflow tract to the aorta has a strong inverse relation to the severity of obstruction. The right parasternal long-axis left ventricular outflow tract view is useful to identify the fibrotic narrowing just proximal to the base of the aortic valve (see Fig. 5). Color-flow Doppler echocardiography reveals turbulent systolic blood flow arising in the left ventricular outflow tract (see Fig. 5). The aortic valve is often mildly thickened, and there is always some degree of aortic insufficiency. The standard method for classification of disease severity is by measurement of peak systolic aortic blood flow velocity using continuous-wave Doppler echocardiography from the left apical five-chamber view or by the subcostal view using a Pedoff probe.
Fig. 5. Echocardiogram of a dog with SAS. (A) Right parasternal short-axis view depicts left ventricular concentric hypertrophy and regions of subendocardial hyperechogenicity consistent with myocardial fibrosis. Short-axis views depict the normal-sized aorta in cross section (B) and the severely stenotic subvalvular fibrotic ring (arrow) immediately below the aorta (C). (D) Discrete fibrotic narrowing (arrow) of the left ventricular outflow tract immediately below the aortic valve is also visualized on the right parasternal long-axis left ventricular outflow tract view. Subendocardial hyperechogenicity and concentric hypertrophy of the interventricular septum and papillary muscle are also seen. (E) Close-up view of the left ventricular outflow tract using color-flow Doppler echocardiographic interrogation shows turbulent systolic blood flow arising at the fibrotic ring in the left ventricular outflow tract. (F) Continuous-wave Doppler echocardiographic measurement of peak systolic aortic blood flow velocity (below the baseline [Ax]) reveals a high velocity of 5.5 m/s and a calculated left ventricular–aortic pressure gradient of 120 mm Hg, confirming the diagnosis of SAS. There is also aortic insufficiency (above the baseline [B+]), with a normal peak diastolic velocity of 4.2 m/s and an aortic–left ventricular diastolic pressure gradient of 70 mm Hg. Ao, aorta; IVS, interventricular septum; LA, left atrium; LV, left ventricle.

(see Fig. 5). The subcostal view yields the highest aortic blood flow velocity, but the difference compared with the left apical view is trivial [5]. Mild, moderate, and severe SAS have been defined as left ventricular outflow tract–aortic pressure gradients of less than 50 mm Hg, 50 to 80 mm Hg, and more than 80 mm Hg, respectively.

The most difficult aspect of diagnosis is distinguishing animals with physiologic murmurs from animals with mild SAS. In mild SAS, there may be a subtle fibrotic ridge in the left ventricular outflow tract and color-flow Doppler echocardiography may show a mild breakup in the laminar signal in that region.
Although normal aortic blood flow velocity is usually 2 m/s or less, there is controversy regarding the highest limit of normal peak aortic blood flow velocity. Boxer dogs frequently have soft systolic left basilar murmurs and aortic blood flow velocities in a range of 2 to 3 m/s. Some investigators have hypothesized that these dogs have an increased sympathetic activation and increased cardiac output that lead to the increased aortic blood flow velocity in the absence of SAS [6]. Because pathologic examination to confirm the presence of mild lesions is not an option, there is an equivocal “gray zone” for the diagnosis of SAS. The presence of aortic insufficiency (in the absence of a VSD, annuloaortic ectasia, or infective endocarditis), turbulent blood flow arising in the left ventricular outflow tract with spectral broadening of the pulsed wave Doppler echocardiographic signal, and mildly elevated velocity greater than 2 m/s support a diagnosis of mild SAS.

Clinical management. Therapy is targeted to prevent sudden death and reduce syncope and exercise intolerance. The mainstay of therapy is the use of beta-blockers (atenolol, 0.5–1.5 mg/kg, administered orally twice daily) to reduce the likelihood of a fatal ventricular arrhythmia. Beta-blockers reduce myocardial oxygen demand, increase coronary perfusion secondary to the negative inotropic and chronotropic effects, and protect the diseased myocardium against arrhythmic effects of sympathetic surges. No studies have compared the effect of beta-blockers versus no treatment on survival. Extrapolation from historical control dogs with severe SAS suggests that dogs treated with atenolol live longer (median survival time of 56 months versus 19 months) [4,7]. Surgical excision of the fibrotic ring and septal myectomy under cardiopulmonary bypass effectively reduce the systolic pressure gradient but do not alter the survival time compared with dogs treated with atenolol [8]. Likewise, balloon valvuloplasty moderately reduces the systolic pressure gradient but
Pulmonic stenosis

PS is the third most common CHD of dogs and is occasionally recognized in cats. PS is caused by a narrowing of the right ventricular outflow tract, pulmonic valve, or main pulmonary artery (subvalvular, valvular, or supravalvular, respectively). Valvular PS is the most common form and consists of variable degrees of valvular thickening, fusion of the cusps, or hypoplasia of the valve annulus. Some valves seem thickened and fused at the commissures but are mobile and doming motion is observed during systole (type 1 dysplasia), whereas others are markedly misshapen, immobile, and severely dysplastic (type 2 dysplasia). Bulldogs and Boxers are predisposed to develop valvular PS or subvalvular PS secondary to an anomalous left coronary artery (R2A type) [9]. A single large coronary artery originating from the right sinus of Valsalva gives rise to the anomalous left coronary artery and the normal right coronary artery. The left coronary artery encircles the pulmonary artery and forms a subvalvular extramural compressive ring.

TVD is a common concurrent defect. Severe right heart failure occurs when there is greater than mild TVD in the face of PS. Using the University of California, Davis, Veterinary Medical Teaching Hospital medical database, approximately 10% of dogs with PS also had a right-to-left shunting patent foramen ovale. If there is significant right-to-left shunting, animals may be hypoxemic, cyanotic, and polycythemic.

PS creates an increased resistance to right ventricular systolic ejection and an elevated right ventricular systolic pressure. Compensatory concentric hypertrophy occurs in an attempt to normalize the increased systolic wall stress. Myocardial fibrosis may occur secondary to severe pressure overload and myocardial ischemia. Right heart failure often occurs if there is concurrent TVD and is likely exacerbated by diastolic right ventricular dysfunction secondary to severe concentric hypertrophy and myocardial fibrosis. Chronic severe PS may also lead to right ventricular myocardial failure.

Dogs with mild PS have a good prognosis; in the absence of concurrent CHD, they should have a normal life expectancy. Sudden death occurred in 30% of dogs with severe valvular PS in one study [10]. During exercise, dogs with severe PS have reduced right coronary artery blood flow and inadequate myocardial perfusion, which may lead to systolic dysfunction, ventricular arrhythmias, and, possibly sudden death [11]. Approximately 35% of dogs with severe PS have clinical signs, including fatigue, syncope, ascites, or cyanosis [10].

The most prominent clinical examination finding is a left basilar holosystolic murmur. If there is concurrent tricuspid regurgitation, there may be a right apical systolic murmur. Femoral pulses are usually good. Right heart failure may be present and consists of jugular venous distention, hepatomegaly, and ascites. Typical electrocardiographic abnormalities include a right axis deviation with
deep S waves of leads I, II, and III as well as the left precordial chest leads. Thoracic radiographs typically show right heart enlargement and a poststenotic dilation of the pulmonary artery (see Fig. 2).

Echocardiography shows right ventricular concentric hypertrophy in proportion to the severity of the obstruction. There may be interventricular septal flattening during systole if the right ventricular systolic pressure exceeds the left ventricular pressure. There is often right atrial enlargement, which may be severe if there is concurrent TVD. The right basilar short-axis and left cranial long-axis views are useful for examination of the pulmonic valve. The pulmonic valve is typically thickened, with fused commissural cusps, and may have variable degrees of mobility with a doming motion during systole (Fig. 6). Color-flow Doppler echocardiography shows turbulent systolic blood flow and is useful to determine the location of the obstruction (see Fig. 6). Pulmonic insufficiency of variable degrees is always present. Continuous-wave Doppler echocardiography is used to determine the severity of the obstruction, with mild, moderate, and severe stenosis defined as a right ventricular outflow tract–pulmonary artery pressure gradient of less than 50 mm Hg, 50 to 80 mm Hg, and greater than 80 mm Hg, respectively (see Fig. 6). Dynamic right ventricular outflow tract obstruction may be seen with infundibular hypertrophy and has a characteristic late systolic peak on continuous wave Doppler echocardiography. Careful examination of the origin and course of the left coronary artery is essential for evaluation of an R2A type anomalous left coronary artery. Measurement of the pulmonary annulus diameter by transthoracic or transesophageal echocardiography is used to determine appropriate balloon diameter if balloon valvuloplasty is planned, and an ideal balloon diameter is 1.2 to 1.4 times the pulmonary artery annulus diameter. A positive-contrast echocardiogram is performed to evaluate for right-to-left shunting defects, such as a patent foramen ovale.

**Clinical management.** Treatment goals are to reduce the systolic right ventricular pressure overload, which may, in turn, reduce the risk of sudden death, reduce clinical signs of syncope or exercise intolerance, reduce right-to-left shunting through a patent foramen ovale, and reduce the severity of tricuspid regurgitation. Balloon valvuloplasty is a relatively noninvasive method to treat severe valvular PS. A right heart catheterization is performed, including pressure measurements and angiography (see Fig. 2). If there is suspicion of an aberrant left coronary artery, left cardiac catheterization and an aortic root angiogram may be needed to evaluate the coronary anatomy. Balloon valvuloplasty is contraindicated in patients with an aberrant left coronary artery. With the aid of a guidewire positioned across the pulmonic valve, a balloon dilation catheter is advanced across the valve and is manually inflated to tear open the stenotic valve (see Fig. 2). Repeat pressure measurements and continuous wave Doppler echocardiographic measurement of the pulmonic blood flow velocity are used to determine the pressure reduction after the valvuloplasty (see Fig. 6). Balloon valvuloplasty is associated with a 53% reduction in the risk of sudden
death in dogs with severe PS and has also been associated with improved quality of life [10]. Balloon valvuloplasty may be considered in less severe PS cases if there is concurrent moderate or severe TVD. Before the use of balloon valvuloplasty, surgical techniques, such as open valvulotomy under inflow

Fig. 6. Echocardiogram of a dog with severe valvular PS. (A) Transesophageal echocardiogram shows the thickened pulmonic valve (arrow) that is fused at the commissures and domes during systole. (B) Color-flow Doppler echocardiography shows turbulent systolic blood flow arising at the opening of the stenotic pulmonic valve. (C) Continuous-wave Doppler echocardiographic measurement of pulmonic blood flow velocity shows severe PS, with a velocity of 5.3 m/s and a right ventricular outflow tract-to-pulmonary artery pressure gradient of 113 mm Hg. After balloon valvuloplasty, the pulmonic valve is able to open during systole (D) and there is laminar systolic blood flow across the valve (E). (F) Continuous-wave Doppler echocardiography after balloon valvuloplasty reveals a markedly reduced right ventricular outflow tract-to-pulmonary artery pressure gradient of 4 mm Hg and pulmonic insufficiency (blood flow velocity below the line). This is an extraordinarily optimal result of balloon valvuloplasty. PA, pulmonary artery; RV, right ventricle.
occlusion or closed valvulotomy via a right ventricular approach, were used to treat valvular PS. Balloon valvuloplasty is not a viable option for the treatment of fibromuscular subvalvular PS or severely dysplastic (type 2) pulmonic valves. Surgical placement of a patch graft at the level of the right ventricular outflow tract and pulmonary artery may be the best option to relieve a muscular obstruction, but the technique confers a high risk of mortality (15%–25% with an experienced cardiac surgeon) or complications [12]. Placement of a valved conduit between the right ventricle and pulmonary artery is a possible treatment option for dogs with aberrant left coronary arteries, but there have been problems with the conduit becoming kinked within the chest cavity.

Medical treatment with beta-blockers has been advocated by some clinicians to minimize the risk of arrhythmic death. Beta-blockers also may be used to minimize a muscular dynamic component of PS after balloon valvuloplasty.

Dysplasia of the Atrioventricular Valves

MVD and TVD are malformations of the mitral valve apparatus or tricuspid valve apparatus, including the valve leaflets, chordae tendineae, or papillary muscles, that result in valvular insufficiency. There are many types of malformations, including short thickened leaflets with clubbed tips, rolling of leaflet edges, short and thick chordae tendineae, fusion of chordae tendineae into a single chord, direct insertion of the papillary muscle to the leaflet, and upward malposition of the papillary muscle causing malalignment of the chordae tendineae [13].

MVD is most commonly seen in large-breed dogs, and Bull Terriers and Great Danes are predisposed. TVD is the most common CHD of Labrador Retrievers and has been shown to be an autosomal dominant inherited trait with incomplete penetrance, localized to chromosome 9 [14]. MVD and TVD are the most common CHDs in cats, and MVD may also be seen in conjunction with atrioventricular canal defects. Thirty-six percent of dogs and 23% of cats with TVD are concurrently affected with MVD [13].

Severe MVD and mitral regurgitation cause elevated left ventricular diastolic pressure, elevated left atrial and pulmonary venous pressures, and the development of pulmonary edema. Congestive heart failure was seen in 75% of dogs with MVD in an early study [13]. Severe TVD and tricuspid regurgitation cause elevated right ventricular end diastolic pressure, elevated right atrial pressure, and right congestive heart failure when right atrial pressure exceeds 10 to 15 mm Hg.

Clinical abnormalities and the pathophysiology of MVD and TVD closely parallel degenerative valve disease. Dogs with MVD commonly have a left apical holosystolic to pansystolic murmur whose intensity usually parallels the severity of the regurgitation. Auscultation of dogs with TVD reveals a right apical systolic murmur in dogs with moderate to severe TVD. In a study of Labrador Retrievers screened by echocardiography for TVD, however, none of the dogs with mild TVD had a murmur, making auscultation an
inappropriate screening test [15]. Dogs with right heart failure may have distended jugular veins, hepatomegaly, and ascites. Electrocardiography may show various supraventricular arrhythmias, such as atrial premature complexes, supraventricular tachycardia, or atrial fibrillation. Splintered QRS complexes (R'R', RR', rR', and rr') are seen in 60% of dogs and cats with TVD [16]. Labrador Retrievers with TVD may also have an accessory pathway bridging the right atrium and the right ventricle, resulting in re-entrant supraventricular tachycardia [17]. Thoracic radiographs demonstrate variable left atrial enlargement, left-sided cardiomegaly, and, possibly, pulmonary venous distention and interstitial-to-alveolar pulmonary infiltrates of the perihilar to caudodorsal lung fields in dogs with MVD. Left-sided congestive heart failure in cats may be characterized by an atypical pattern of edema distribution or pleural effusion. Dogs with moderate to severe TVD often have profound right atrial dilation as well as a dilated caudal vena cava and hepatomegaly (see Fig. 4). Cats with TVD have right atrial enlargement and a dilated and often tortuous caudal vena cava. Cats with right heart failure may develop pleural effusion with or without ascites.

Echocardiography reveals morphologic abnormalities of the mitral valve apparatus or tricuspid valve apparatus, as described previously. There may be left atrial enlargement, eccentric left ventricular hypertrophy secondary to volume overload, and, possibly, mild myocardial failure secondary to chronic volume overload in medium- to large-breed dogs and cats with MVD. TVD may cause profound right atrial dilation that may distort the position of the other chambers, making it difficult to obtain normal standard views (Fig. 7). Color-flow Doppler echocardiography is useful to identify the presence of valvular regurgitation (see Fig. 7).

Clinical management. Medical management of congestive heart failure with furosemide (1–4 mg/kg administered orally every 24 hours to three times daily) and an angiotensin-converting enzyme (ACE) inhibitor is standard therapy. Treatment of tachyarrhythmias with antiarrhythmics may also be necessary. Intracardiac and extracardiac annuloplasty has been performed to reduce the severity of valvular insufficiency. Surgical valve repair or replacement during cardiopulmonary bypass has been successfully performed by a few highly experienced cardiac surgeons, but there is a high risk of mortality, and this procedure is limited to medium- and large-breed fully grown dogs. Dogs must remain on lifelong coumarin anticoagulant therapy [18].

Mitral stenosis and tricuspid stenosis

Stenosis of the atrioventricular valves is rarely seen in dogs and cats; if present, it occurs in conjunction with valve dysplasia and regurgitation. Atrioventricular valve stenosis is caused by narrowing of the valve orifice, which impedes atrioventricular filling and results in elevated atrial pressure. Mitral stenosis is seen in Newfoundlands and Bull Terriers. SAS is a common concurrent defect in Newfoundlands. Mitral stenosis commonly leads to left-sided congestive heart failure in almost 75% of dogs [19]. Pulmonary hypertension may occur as
a result of hypoxic pulmonary arteriolar vasoconstriction and increased pulmonary venous pressure. Supravalvular mitral stenosis is a rare defect reported in cats, where a fibrotic ring or perforated membrane separates the left atrium into two chambers, with the obstructed proximal chamber usually including the auricle.

Auscultatory findings commonly include an apical systolic murmur in more than 50% of cases because of concurrent atrioventricular valvular insufficiency and, less commonly, a soft diastolic murmur in only 27% of dogs with mitral stenosis [19].

Echocardiography is necessary to diagnose mitral or tricuspid stenosis. Two-dimensional views show a restricted immobile mitral or tricuspid valve with lack of diastolic separation of the leaflets. M-mode across the mitral valve using a right parasternal short-axis view in animals with mitral stenosis reveals decreased mitral leaflet separation during diastole and a reduced E-to-F slope [19]. Color-flow Doppler echocardiography shows aliasing to turbulent mitral or tricuspid diastolic inflow and often also identifies systolic valvular insufficiency. Pulsed-wave or continuous-wave Doppler echocardiography across the affected valve shows increased peak inflow velocity and prolonged pressure half-times. The maximum atrial-to-ventricular pressure gradient is increased and may be used to quantify atrial pressure noninvasively.

The prognosis is poor for animals with significant mitral or tricuspid stenosis. In one study, 60% of dogs with mitral stenosis died by the age of 2.5 years [19]. Medical treatment consists of diuretics and ACE inhibitors, but aggressive diuresis must be avoided, because there is reliance on an increased atrial pressure for ventricular filling across the stenotic valve. Tachycardia can worsen diastolic filling and result in higher atrial pressure and reduced cardiac output, and antiarrhythmic therapy may be needed to treat tachyarrhythmias. Balloon
valvuloplasty has been performed on several dogs with tricuspid stenosis, with successful reduction of pressure gradients and clinical improvement [20]. Surgical options include an open or closed commissurotomy. Supravalvular mitral stenosis may be surgically excised under inflow occlusion or during open-heart surgery under cardiopulmonary bypass.

Lesions Causing Systemic-to-Pulmonary Shunting

Patent ductus arteriosus

PDA is one of the two most common CHDs in dogs and is rare in cats. In the fetus, the ductus arteriosus connects the pulmonary circulation with the systemic circulation to bypass the nonfunctional lungs. Approximately 80% to 90% of the right ventricular output is diverted from the pulmonary artery through the ductus to the aorta. At birth, lungs inflate, the pulmonary vascular resistance drops to 20% of the systemic vascular resistance, and the blood flow reverses. Oxygenated arterial blood in the ductus inhibits prostaglandin release and triggers the circumferentially oriented smooth muscle cells of the ductus to contract. The ductus arteriosus closes within the first 2 days of life and is securely closed after 7 to 10 days [21]. The defect responsible for a PDA is hypoplasia of the smooth muscle fibers of the ductus arteriosus [21]. PDA morphology has been divided into six grades depending on the amount of smooth muscle mass within the ductus [21]. The amount of smooth muscle cells within the ductus determines the shape of the ductus, and therefore the amount of the shunt. Nearly all ducts are funnel shaped, with the greatest amount of smooth muscle cells and constriction at the pulmonary artery end. Higher grades have less smooth muscle and less tapering at the pulmonary artery end. A grade 6 ductus almost completely lacks smooth muscle, resulting in a wide-open conduit and a reverse PDA physiology, which is discussed in the section on pulmonary-to-systemic shunting.

The amount of blood shunted across the PDA is related to the size of the ductal opening and the resistance of the pulmonary and systemic arterial beds. In left-to-right shunts, the shunt circuit includes the aorta up to the level of the ductus, ductus, pulmonary arteries, pulmonary veins, left atrium, and left ventricle. In addition to the normal cardiac output, the structures within the shunt circuit must encompass the extra shunt volume, which may be quite large in a grade 4 or 5 ductus. The result is enlargement of chambers and vessels included in the shunt circuit. If there is a large left-to-right shunt, there is elevated left ventricular end diastolic pressure, left atrial pressure, and pulmonary venous pressure, which may lead to congestive heart failure. Rarely, if there is severe volume overload of the pulmonary circulation, the pulmonary arterioles may hypertrophy and pulmonary hypertension may develop, causing the left-to-right shunt to diminish and possibly reverse to a right-to-left shunt. The window of time to repair a PDA is before development of pulmonary hypertension, because PDA closure in the face of severe pulmonary hypertension is contraindicated.
PDA is heritable in Poodles as a polygenic or threshold trait, with sex as a modifying factor (ie, female predisposition). Other breeds commonly affected are listed in Table 1. Auscultation of a PDA reveals the pathognomonic continuous murmur located in the left axillary region. If auscultation is performed too caudally at the apex, the continuous murmur may be missed and the murmur may be incorrectly classified as systolic. Femoral pulses are hyperkinetic (ie, bounding) because of the mild increase in aortic systolic pressure and the markedly reduced diastolic pressure caused by the continued diastolic shunting to the pulmonary circulation. If there is a large left-to-right shunt, the animal may be dyspneic and there may be adventitial pulmonary sounds on auscultation. Thoracic radiographs often reveal left-sided cardiomegaly and a ductal aneurysm (ie, ductal bump), which is the dilation of the aorta and the aortic region of the PDA proximal to the insertion to the pulmonary artery (see Fig. 3). An overcirculation pattern of the pulmonary vasculature may be seen. Evaluation of the lungs for evidence of congestive heart failure is essential for optimal stabilization before closure of the PDA.

Echocardiography is necessary to confirm the diagnosis, evaluate for other concurrent CHDs, and assess whether the patient is a suitable candidate for surgical or coil embolization closure of the PDA. The left ventricle appears eccentrically hypertrophied as a result of volume overload. Often, there is myocardial failure, as evidenced by an increased end systolic diameter and increased E point–to-septal separation (EPSS) of the mitral valve during early diastole. EPSS is increased with systolic myocardial failure, because there is an increased volume of blood remaining within the left ventricle at the end of systole, causing a lower left atrial–to–left ventricular pressure gradient during early diastole and reduced early diastolic flow. Less flow results in reduced mitral valve excursion and can be identified by M-mode measurement of the EPSS. Usually, fractional shortening remains normal. Myocardial failure is more common and more severe in older animals. Left atrial enlargement is generally mild to moderate. Visualization of the ductus is possible by obtaining the right parasternal short-axis basilar view and, preferably, the left cranial parasternal long-axis view of the pulmonary artery. Measurements of the minimal ductal opening size at the insertion to the pulmonary artery and maximal ductal diameter can be made by transthoracic or transesophageal echocardiography to assess whether the dog is a good candidate for coil embolization (Fig. 8). Color-flow Doppler echocardiography at the level of the pulmonary artery and ductus shows continuous turbulent blood flow shunting into the pulmonary artery from the ductus as well as laminar ductal flow (see Fig. 8). Continuous-wave Doppler echocardiography is aligned parallel to the turbulent pulmonary arterial blood flow to measure peak systolic velocity and calculate the aortic-to-pulmonary artery pressure gradient. This is useful to evaluate for pulmonary hypertension, which would result in a lower pressure gradient than expected in a normotensive animal (ie, <4.5 m/s). Centrally arising functional mitral regurgitation is often seen on color-flow Doppler echocardiography and occurs secondary to left ventricular eccentric hypertrophy and mitral annular dilation.
Peak aortic systolic blood flow velocity is often mildly increased (ie, 1.8–2.5 m/s) because of the increased left ventricular stroke volume.

**Clinical management.** Treatment of congestive heart failure with diuretics is essential before surgery or coil embolization. Correction of a PDA should be performed as soon as reasonably possible to avoid the development of congestive heart failure or pulmonary hypertension. In the past, the only option to close a PDA was surgical ligation. In the hands of an experienced surgeon who has performed more than 25 PDA ligations, the complication rate is low, at less than 5% [21]. There may be silent residual flow detected by color-flow Doppler echocardiography through the ductus after the ligation procedure in 20% of dogs. Less than 2% of dogs develop recanalization that requires repeat ligation, however. Echocardiography after ductal closure typically shows a reduced left ventricular diastolic diameter, unchanged end systolic diameter, and moderately decreased fractional shortening of 18% to 25%. The left atrium is smaller and often normal in size. The ductal aneurysm usually persists, and there may be some degree of left-sided cardiomegaly on thoracic radiographs. The
prognosis after ductal ligation is good. The prognosis without ductal closure is grave, and in one study, 64% of puppies died before 1 year of age [22]. Surgical closure is the only option for cats with left-to-right shunting PDAs.

Percutaneous transarterial coil embolization has become an attractive less invasive option for closure of small to moderately sized PDAs in dogs. The femoral artery is catheterized, and an aortic root angiogram is performed to identify the PDA (see Fig. 3). Under fluoroscopic guidance, a thrombogenic stainless-steel coil with Dacron fibers is positioned within the ductus at the narrowed region near the insertion to the pulmonary artery. Several coils are often placed to achieve adequate closure of the ductus. Angiography and transesophageal echocardiography are used to evaluate the amount of closure of the PDA (see Figs. 3 and 8). In extremely small animals with moderate shunts, the carotid approach may be used for coil embolization. Of the last 125 cases of attempted coil embolization at the University of California, Davis, 86% of cases had successful coil implantation, with a low mortality rate of 2.4% [23]. Aberrant coil embolization of a single coil to the pulmonary artery is tolerated without abnormalities in ventilation perfusion scans [24]. Retrieval of coils that are aberrantly embolized to the aorta or femoral artery is recommended. A considerable number of dogs may have hemodynamically insignificant residual ductal flow seen on echocardiography after coil embolization, which often reduces over the following months [23].

**Ventricular septal defect**

A VSD is an uncommon CHD in dogs and is one of the most common CHDs in cats [25]. VSDs can occur in several regions of the heart, most commonly in the basilar perimembranous region just below the right coronary and noncoronary aortic cusps on the left side and below the septal leaflet of the tricuspid valve on the right. Muscular VSDs are uncommon. Because the defect usually lies just beneath the aortic valve, there may be a lack of support of the aortic valve, resulting in mild dextroposition of the aorta and aortic insufficiency. Shunting across the defect depends on the size of the communication as well as the pulmonary and systemic vascular resistances. Small defects are resistive to flow, resulting in predominantly systolic shunting of flow from the left ventricle to the right ventricle. The added shunt volume is immediately ejected out to the pulmonary artery, avoiding volume overload of the right heart. The chambers and vasculature that are affected by the increased blood volume include the left ventricle, pulmonary artery, pulmonary veins, and left atrium. Congestive heart failure occurs if there is a moderate to severe left-to-right shunt. A large VSD with nonresistive left-to-right flow may lead to the development of Eisenmenger’s syndrome, which is discussed in the section on pulmonary-to-systemic shunting defects.

Conotruncal malformations, including VSD and TOF, have been found to be recessively inherited in Keeshonds. In English Springer Spaniels, the heritability is autosomal dominant with incomplete penetrance or attributable to a polygenic mechanism. The cause of a VSD is unknown but likely involves
lack of fusion of the muscular septum with the endocardial cushions or malfor-
mation of the conal septum. The typical auscultation finding is a loud holosys-
tolic right basilar murmur. The murmur is not continuous, because there is
only systolic shunting. The murmur intensity in an animal with a large VSD
and some degree of pulmonary hypertension is softer, because there is a lower
pressure gradient and velocity of shunted blood. Femoral pulses are normal.
Thoracic radiographs may be normal if the VSD is small. If there is a moderate
to large shunt, there may be pulmonary overcirculation and left atrial and ven-
tricular enlargement.

Echocardiography is useful to confirm the diagnosis and to assess the sever-
ity of the shunt. In a small shunt, heart size is normal and careful inspection of
the left ventricular outflow tract in the region of the membranous septum may
reveal a small defect. Often, small defects are difficult to visualize on two-
dimensional echocardiography, and color-flow Doppler echocardiographic in-
terrogation of the basilar interventricular septum, including the right ventricu-
lar side, is needed to identify the turbulent systolic blood flow shunting from
left to right. Continuous-wave Doppler echocardiography is used to measure
the velocity of the shunt and to calculate the left ventricular–to–right ventricu-
lar pressure gradient. Knowing that the peak left ventricular systolic pressure
is approximately 120 mm Hg and the peak right ventricular systolic pressure
is approximately 20 mm Hg in a normal animal, the pressure gradient is
100 mm Hg. The peak systolic velocity should be greater than 4.5 m/s (pres-
ture gradient of 80 mm Hg) in a small to moderate left-to-right VSD. Pulmo-
nary hypertension is present if the peak VSD velocity is less than 4.5 m/s in
a normotensive animal.

Atrial septal defect
ASDs are rare in dogs and uncommon in cats. Embryologic formation of the
atrial septum occurs by growth of two atrial membranes. The primum septum
grows from the roof the atria to the floor of the atria. The secundum septum
grows from the roof of the atria on the right side of the primum septum. A de-
fect in the development of the primum atrial septum results in an ostium low in
the atria, below the foramen ovale, near the atrioventricular valves. Ostium pri-
num ASDs are most commonly seen in cats. Atrioventricular canal defects are
endocardial cushion defects that include an ostium primum ASD and a VSD.
Abnormal development of the septum secundum results in an ostium at or
above the level of the foramen ovale. Ostium secundum ASDs are most com-
monly seen in dogs.

Physical examination in animals with moderate ASDs reveals a grade II to
III/VI left basilar holosystolic murmur that is created as a result of functional
PS from the large right ventricular stroke volume. A split S2 heart sound
may be present. Thoracic radiographs reveal right- and left-sided cardiomegaly,
pulmonary overcirculation, and, possibly, pulmonary edema in large shunts.
Echocardiography is necessary to determine the diagnosis and severity of the
left-to-right shunt. The right atrium and right ventricle are dilated, often to
a greater degree than the left atrium and left ventricle. An ASD must be visualized with the ultrasound beam perpendicular to the interatrial septum (ie, the right parasternal long-axis view); otherwise, an erroneous diagnosis might be made based on an artifact of septal fallout. Color-flow Doppler echocardiography is necessary to visualize left-to-right shunting blood across the defect, and pulsed-wave Doppler echocardiography helps to confirm the low velocity of shunting blood. A positive contrast bubblegram can also be used to identify a negative contrast jet of anechoic blood entering the opacified right atrium. The pulmonary artery blood flow velocity is mildly increased.

Clinical management. Treatment of small septal defects is not necessary, because there is a normal lifespan with small shunts. Definitive treatment of large left-to-right shunting defects is patch closure under cardiopulmonary bypass [26]. This technique is limited to only a few surgical centers in the country and is not an option for cats or extremely small dogs. An alternative to patch closure in animals with large left-to-right shunting septal defects and congestive heart failure is pulmonary artery banding. Pulmonary artery banding is used to increase the pulmonary artery resistance and reduce the magnitude of the left-to-right shunt [26]. This procedure should not be performed in animals less than 6 months of age, because the band becomes too tight as the animal grows and results in a right-to-left shunt or right ventricular failure. The band is constricted until the pulmonary artery diameter is one third the diameter of a normal pulmonary artery. Several suggested guidelines for intraoperative evaluation of the optimal degree of banding include a 50% reduction in pulmonary artery pressure and a 50% to 100% increase in right ventricular pressure [26]. Intraoperative echocardiography may be useful for monitoring the degree and direction of the shunt and left ventricular–to–right ventricular pressure gradient of a VSD. Medical treatment consists of systemic arterial vasodilation with amlodipine or hydralazine to reduce the severity of the left-to-right shunt. Palliative medical therapy with diuretics and ACE inhibitors is used for treatment of congestive heart failure. Positive inotropic drugs may be needed if there is myocardial failure, often as a result of severe aortic insufficiency.

Lesions Causing Pulmonary-to-Systemic Shunting and Cyanosis
tetralogy of Fallot
TOF is the most common CHD causing cyanosis. Components of TOF include PS, concentric right ventricular hypertrophy secondary to the pressure overload, a subaortic VSD, and an overriding dextropositioned aorta. TOF is caused by abnormal embryonic development of the conotruncal septum, resulting in varying degrees of infundibular and valvular PS, pulmonary artery hypoplasia, malalignment of the infundibular septum, and a VSD. TOF is uncommon in the dog and cat. Genetic studies have defined TOF as a simple autosomal recessive inherited trait in Keeshonds. Other common concurrent CHDs include a PDA, an ASD, and aortic arch defects. There may be other concurrent congenital defects, including tracheal hypoplasia, peritoneal
diaphragmatic defect, pectus excavatum, retinal dysplasia, and persistent pupillary membranes. The severity of the PS and the systemic vascular resistance determine the severity of right-to-left shunting. Right-to-left shunting results in arterial hypoxemia and triggers erythropoietin production by the kidneys. The PaO₂ is usually 35 to 40 mm Hg in cyanotic animals. Moderate polycythemia (55%–65% packed cell volume [PCV]) is beneficial, but a hyperviscosity syndrome may occur when the PCV exceeds 70%. Bronchoesophageal arteries often hypertrophy to provide blood flow to the lungs in severe cases.

Murmur severity inversely correlates with the severity of the PS. For example, if there is mild PS and a small VSD, there may be a loud right basilar holosystolic murmur caused by the VSD shunt. Conversely, if there is severe PS, a large VSD, and a hyperviscosity syndrome, a murmur may be absent. Thoracic radiographs show right ventricular enlargement and often demonstrate pulmonary vascular undercirculation. A right ventricular hypertrophy pattern is seen on the electrocardiogram. Echocardiography reveals concentric right ventricular hypertrophy, PS (often a combination of infundibular and valvular), and, often, pulmonary artery hypoplasia. Using the right parasternal long-axis left ventricular outflow tract view, the VSD is visualized in the perimembranous subaortic region and there is a dextrapositioned aorta overriding the ventricular septum. Color-flow Doppler echocardiography is used to visualize laminar right-to-left shunting flow from the right ventricle to the aorta. A positive-contrast echocardiogram is also useful to demonstrate the degree of right-to-left shunting. Continuous-wave Doppler echocardiography is used to measure the velocity of the pulmonic blood flow and to calculate the PS pressure gradient. In the presence of a large VSD, however, the pressure gradient is not especially helpful for determination of severity of the PS because it mirrors the left ventricular–to-aortic pressure gradient (usually ~100 mm Hg).

**Right-to-left patent ductus arteriosus**

A right-to-left PDA is a rare CHD that is a result of large nonresistive grade 6 ductal morphology with virtually no ductal smooth muscle. Because there is a large communication between the pulmonary and systemic vascular beds, the pulmonic and systemic vascular resistances and pressures equilibrate. Irreversible pulmonary arteriolar changes occur, including medial and intimal smooth muscle hypertrophy and fibrosis, obliteration of pulmonary arteries with plexiform lesions, and microvascular thrombosis. The severely increased pulmonary vascular resistance becomes fixed and is higher than the systemic vascular resistance. Unoxygenated blood flow shunts from the pulmonary artery to the descending aorta and results in differential cyanosis of the caudal half of the body. Renal hypoxia results in erythropoietin production and secondary polycythemia. Animals with a large left-to-right shunting PDA (ie, grade 5) may develop Eisenmenger’s syndrome and reversal of the PDA shunt.

Animals with a right-to-left PDA do not have a murmur, because there is laminar right-to-left or bidirectional ductal flow. There may be a split S₂ heart
sound secondary to severe pulmonary hypertension. Thoracic radiographs reveal right heart enlargement, a ductal aneurysm of the aorta, and pulmonary vascular undercirculation. Electrocardiography shows a right ventricular hypertrophy pattern. Echocardiography shows severe right ventricular concentric hypertrophy, interventricular septal flattening, and pulmonary artery dilation. A large nontapered PDA may be visualized using the right parasternal short-axis basilar view or the left cranial parasternal long-axis view. Color-flow Doppler echocardiography shows laminar right-to-left or bidirectional shunting blood flow within the ductus. Care must be taken to avoid the incorrect classification of a normal left main pulmonary artery as a right-to-left shunting PDA in an animal with pulmonary hypertension, however. A positive-contrast echocardiogram is necessary to confirm the diagnosis of a PDA. Visualization of bubbles within the abdominal aorta in the absence of an intracardiac right-to-left shunt confirms the diagnosis.

**Eisenmenger’s syndrome**

Eisenmenger’s syndrome occurs when there is a large nonresistive defect (PDA, VSD, ASD, or atrioventricular canal defect) with an initial large left-to-right shunt that causes pulmonary arterial hypertension and ultimately results in right-to-left shunting. Initially, the pulmonic blood flow is greatly increased because of the large left-to-right shunt. Over time, the pulmonary arterioles develop irreversible pathologic changes that result in a severely elevated and fixed pulmonary vascular resistance. Blood flow then reverses to shunt from right to left, because systemic vascular resistance is lower than pulmonic vascular resistance. This results in arterial hypoxemia, cyanosis, and secondary polycythemia.

**Clinical management of cyanotic heart defects**

Open surgical repair of TOF under cardiopulmonary bypass is a feasible option but has rarely been done in veterinary medicine. Surgical correction of a right-to-left shunting PDA or closure of defects in patients with Eisenmenger’s syndrome is contraindicated. Avoidance of systemic hypotension is essential in animals with right-to-left shunting defects so as to avoid worsened hypoxemia. Medical management is aimed at controlling the severity of polycythemia and avoidance of a hyperviscosity syndrome. The goal of medical management is aimed at maintenance of a PCV at 60% to 68%. Phlebotomy is a useful and safe technique to reduce polycythemia. The amount of blood to be removed can be calculated using the following formula:

\[
\text{blood to be removed (mL)} = \left[ \text{body weight (kg)} \times 0.08 \right] \times 1000 \text{ mL/Kg} \\
\times \left[ \left( \frac{\text{actual hematocrit} - \text{desired hematocrit}}{\text{actual hematocrit}} \right) \right].
\]

The amount of blood removed is simultaneously replaced with one to two times the volume of intravenous fluids. Another alternative is removal of 10% of the blood volume in the morning without fluid replacement and
removal of 2% to 10% of the blood volume in the afternoon. If phlebotomies are poorly tolerated or must be performed too frequently, reversible bone marrow suppression of red blood cell production using hydroxyurea is another treatment option. The initial loading dose of 30 mg/kg/d is given for 1 week, followed by a maintenance dose of 15 mg/kg/d. A CBC and platelet count must be performed initially every 1 to 2 weeks. Side effects of the medication include vomiting, diarrhea, anorexia, bone marrow hypoplasia, and toenail soughing. Survival is variable in animals with cyanotic heart disease and often ranges from 1 to 5 years.

OTHER CONGENITAL HEART DISEASES

Cor Triatriatum

Cor triatriatum dexter is a rare CHD caused by persistence of the right sinus venosus valve and results in a persistent membrane that partitions the right atrium into a cranial (normal) chamber and an obstructed caudal chamber. The caudal vena cava drains into the caudal obstructed chamber, and the cranial vena cava and azygous vein drain into the normal cranial chamber of the right atrium. This results in severe ascites, hepatomegaly, and hepatic venous distention in the absence of jugular venous distention or pleural effusion. This defect has only been described in dogs, and there is no breed predisposition. The only successful treatment of cor triatriatum dexter is to reduce the obstruction of the membrane manually by surgical excision under inflow occlusion or, possibly, by percutaneous balloon dilation. Cor triatriatum sinister is caused by lack of normal regression of the fetal pulmonary veins to form the roof of the left atrium and results in an obstructive membrane dividing the left atrium into a caudodorsal obstructed chamber and a cranial nonobstructed chamber. The left auricle is located within the nonobstructed chamber. This aids in distinguishing cor triatriatum sinister from supravalvular mitral stenosis. Cor triatriatum sinister has been described in cats and people. Obstruction of pulmonary venous return into the left atrium results in severe left-sided congestive heart failure. Surgical excision of the obstructive membrane under inflow occlusion or during cardiopulmonary bypass is necessary to relieve the obstruction.

Vascular Ring Anomalies

A persistent right aortic arch (PRAA) is the most common vascular ring anomaly and is commonly seen in German Shepherd Dogs and Irish Setters. Normally, the right fourth aortic arch transforms in utero to form the brachiocephalic trunk and right subclavian artery, and the left fourth aortic arch remains as the adult aortic arch. In animals with PRAA, the right aortic arch is connected to the (left) pulmonary artery by the left-sided PDA, which courses over the esophagus and trachea. After birth, the PDA closes and becomes the ligamentum arteriosum, which compresses the esophagus or trachea. A PRAA does not result in cardiovascular abnormalities. Fifty percent of dogs with a PRAA also have a persistent left cranial vena cava. A PRAA
often becomes clinically evident in weanlings that are transitioning to a solid food diet. Other vascular ring abnormalities are rare (<5%) and include a double aortic arch, retroesophageal left subclavian artery with a right-sided ligamentum arteriosum, or left aortic arch with a right-sided ligamentum arteriosum. Moderate or marked focal leftward curvature of the trachea near the cranial border of the heart in dorsoventral or ventrodorsal radiographs is a consistent abnormality in animals with vascular ring abnormalities and is not seen in animals with generalized megaesophagus [27]. Barium esophagograms may also be useful to distinguish vascular ring abnormalities from generalized megaesophagus, but misdiagnosis may occur if there is inadequate distal esophageal filling. Treatment of a PRAA is surgical excision of the ligamentum arteriosus and division of a retroesophageal subclavian artery if present. Early surgical treatment is recommended to avoid irreversible damage to the esophagus. The prognosis with early surgical treatment is fair, and dietary management is also needed.

**Peritoneal Pericardial Diaphragmatic Hernia**

A peritoneal pericardial diaphragmatic hernia (PPDH) is the most common congenital pericardial defect in dogs and cats. A PPDA is caused by a defect in the ventral diaphragm and pericardium that allows contents of the abdominal cavity to enter the pericardial cavity. Liver lobes and the omentum are the most common herniated structures, although the intestines, stomach, or spleen may also become herniated. A PPDH is more common in cats than in dogs, and Persians and Himalayans are predisposed. Most animals are asymptomatic, and the diagnosis is often made incidentally by identification of severe cardiomegaly with differential intrapericardial opacities, possibly including gas-filled bowel loops. In symptomatic animals, the most common clinical signs are tachypnea, dyspnea, vomiting, and anorexia. Concurrent sternal malformations, such as a pectus excavatum, an incomplete xiphoid, and fused sternebrae, are relatively common in dogs and uncommon in cats. Surgical repair is indicated in symptomatic animals.

**SUMMARY OF CONGENITAL HEART DISEASES**

Congenital heart defects are divided into the following main categories:

- Defects causing pressure overload
  - SAS and PS
- Defects causing volume overload
  - MVD and TVD
- Systemic-to-pulmonary shunts
  - PDA, ASD, and VSD
- Defects causing cyanosis from pulmonary-to-systemic shunts
  - TOF, right-to-left PDA, and Eisenmenger’s syndrome

The clinical diagnostic workup of CHD includes proper localization and characterization of the murmur, thoracic radiographs, and electrocardiography,
and a definitive diagnosis requires echocardiography. PDA and valvular PS are CHDs that may be treated successfully by surgical or interventional catheter-based procedures. Furosemide and ACE inhibitors are used for palliative treatment of congestive heart failure. Medical management of patients with cyanotic CHD consists of control of polycythemia, with the hematocrit maintained between 60% and 68%.

References


