HEART OF THE MATTER
When assessing a patient with a congenital heart defect, it can be useful to determine which type of load on the heart has been altered and which chamber is affected.
A Look at Unusual Congenital Heart Defects in Dogs and Cats

CONGENITAL HEART DEFECTS (CHDs) are well-recognized causes of morbidity and mortality in veterinary medicine (BOX 1). Yet, in comparison to all other illnesses, they account for a very small percentage of morbidity, with a 1999 reported incidence in dogs of 0.46% to 0.85%. A search of the Veterinary Medical Database (maintained by a consortium of veterinary medicine colleges) for any diagnosis of CHD in all dogs entered between 1965 and 2003 yielded a calculated incidence of 0.9%, while a 2015 report put the incidence of CHDs at 0.13% in mixed-breed dogs and 0.14% in mixed-breed cats. The lower percentage for dogs in this report is thought to be due to the lack of purebred dogs in the study group, not a true decrease in the incidence of CHDs. This article explores the rarest CHDs, which present an opportunity to better understand cardiac physiology.

PHYSIOLOGY REVIEW

Blood pressure is the product of cardiac output and systemic vascular resistance (FIGURE 1). Systemic vascular resistance is controlled by the arterioles via vasodilation or constriction, while cardiac output is the product of the stroke volume and heart rate. The stroke volume is determined by 3 influences: preload, afterload, and contractility. Very simply, preload can be thought of as the volume ready to fill the ventricle, whereas afterload represents the force (pressure) the ventricle must exert to move blood forward. Contractility is usually thought of as the ability of the...
myocardium to perform its work. Therefore, preload is a volume load, and afterload is a pressure load.

When assessing a patient with a CHD, it can be useful to determine which load on the heart has been altered and which chamber is affected. For example, pulmonic stenosis (PS) restricts blood flow out of the right ventricle (RV), which increases the pressure load (afterload) on the RV, resulting in hypertrophy. This increased pressure in the RV, right atrium (RA), and caudal venous system can lead to signs of right-sided congestive heart failure (CHF), such as ascites, organomegaly, anorexia, cachexia, and dyspnea. Left-sided CHF includes failure of the left atrium (LA) and left ventricle (LV) and leads to pulmonary edema.

Any CHD can, therefore, be assessed by starting with the fundamental physiology and working backward to predict the outcome. To aid in this assessment, knowledge of the normal pressure of each heart chamber is helpful (TABLE 1). Blood always flows from an area of high pressure to one of low pressure and takes the path of least resistance.

Patients can have more than one form of CHD. The author has examined a dog with tetralogy of Fallot (TOF) and patent ductus arteriosus (PDA). These combinations sometimes present a mechanism for compensation that allows the patient to live relatively normally as a modern pet. One consequence of TOF is reduced pulmonary perfusion or a mismatch in ventilation-perfusion equity. In the patient mentioned above, the PDA allowed extra blood to shunt to the lungs, thereby maintaining the ventilation-perfusion equity closer to normal.

### ATRIOVENTRICULAR VALVE DYSPLASIA

Valve dysplasia is abnormal development of the heart valves. Common examples are PS and subaortic stenosis. Tricuspid valve dysplasia (TVD), mitral valve dysplasia, and cor triatriatum dexter and sinister are, respectively, less common.

### Tricuspid Valve Dysplasia

TVD is a malformation of the tricuspid valve (TV); the effect may be inconsequential or cause serious morbidity. Labrador retrievers are overrepresented for this disease.

A dysplastic TV may have fused and immobile leaflets with a stenotic orifice or elongated leaflets that are...
incompetent. TV displacement into the RV is known as Ebstein’s anomaly. TVD may cause regurgitation and/or stenosis, thereby increasing volume and pressure on the RA and leading to right-sided CHF.

Clinical Signs and Physical Examination Findings
TVD is typically diagnosed in young adult or middle-aged patients; some cats may develop CHF as kittens. Presenting complaints include exercise intolerance, lethargy, syncope, and abdominal distention.

On physical examination, a murmur may or may not be present. If present, it is typically loudest on the right hemithorax, is holosystolic with variable intensity, and may have a crescendo quality. Jugular distention occurs with significantly elevated RA pressure, and jugular pulses occur with significant tricuspid regurgitation. A hepatojugular reflux test can be performed by gently lifting the cranial abdomen while observing the jugular veins, displacing some blood toward the RA and the cranial vena cava. A wave of jugular distention noted after the abdominal lift is a positive result indicating elevated RA pressure.

Diagnostic Tests
Radiography, echocardiography, and electrocardiography (ECG) are used to diagnose TVD if an arrhythmia is present during the physical examination. Thoracic radiographs reveal an enlarged cardiac silhouette in the RA and RV region. The heart may appear somewhat globoid if RA enlargement is severe. The caudal vena cava will be enlarged. The ventrodorsal radiograph often shows a characteristic “reverse D” appearance over the right heart chambers. Enlargement of the RA and RV may be extreme. Ascites can often be noted in the cranial abdomen on the thoracic radiograph.

ECG generally shows a normal sinus rhythm or sinus tachycardia unless disease progression is advanced, when arrhythmias such as atrial fibrillation or ventricular premature complexes may be present. The mean electrical axis (MEA) deviates to the right, and the QRS complex may be “splintered” due to a ventricular conduction system defect (FIGURE 2).

Typical findings on an echocardiogram include a dilated RA, which may be several times the normal size (FIGURE 3). The TV can be visualized for movement. Tethering and poorly mobile leaflets are common. Redundant chordae tendineae may be seen. Dilation of the RV is also common. The LA and LV are often normal in size or slightly diminished depending on RV output. Doppler echocardiography can be used to visualize regurgitation, measure high-velocity flow related to stenosis, and estimate pulmonary artery (PA) pressure by measuring tricuspid regurgitation velocity. Patent foramen ovale (PFO) may be noted by Doppler imaging and confirmed with a contrast echocardiogram.

<table>
<thead>
<tr>
<th></th>
<th>SYSTOLE (MM HG)</th>
<th>DIASTOLE (MM HG)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right atrium</td>
<td>4–6</td>
<td>0–4</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>15–30</td>
<td>5–15</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>15–30</td>
<td>5–15</td>
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<tr>
<td>Pulmonary wedge</td>
<td>6–12</td>
<td>4–8</td>
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<tr>
<td>Left atrium</td>
<td>5–12</td>
<td>&lt;8</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>95–150</td>
<td>&lt;10</td>
</tr>
<tr>
<td>Aorta</td>
<td>95–150</td>
<td>70–100</td>
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FIGURE 2. Electrocardiogram from a dog with TVD. The heart rate is 120 beats per minute. The rhythm is sinus arrhythmia, with an MEA of approximately −105° on the hexaxial system. The QRS complex is more isoelectric, with nearly equal distribution of the Q and the R waves, creating a “splintering” effect. This is characteristic of ventricular conduction defects common with TVD.
Management
Management is primarily control of signs of right-sided CHF. Diuretics, angiotensin-converting enzyme inhibitors, inodilators, and spironolactone are the mainstays of therapy. If the valve is stenotic, balloon valvuloplasty has been successful in palliative therapy; however, clinical signs may return owing to increased tricuspid regurgitation. Surgical replacement or repair of the TV may be done via cardiopulmonary bypass.

Mitral Valve Dysplasia
Mitral valve dysplasia is very rare. It is very similar to tricuspid regurgitation. The valve may be stenotic, incompetent, or both. Stenosis is less prevalent than insufficient valves, which demonstrate a wide range of deformities.

Clinical Signs and Physical Examination Findings
These patients present with signs of left-sided CHF, including dyspnea, tachypnea, weakness, lethargy, exercise intolerance, and possibly syncope. During physical examination, cyanosis, weak femoral pulses, pulmonary crackles, coughing (in dogs), and thoracic fluid lines (in cats) may be appreciated. Both systolic and/or diastolic murmurs may be heard.

Diagnostic Tests
Radiographs show an enlarged cardiac silhouette in the caudal heart over the LA and dilation of the pulmonary veins. Infiltrates consistent with pulmonary edema may be present, indicating left-sided CHF. Some dilation of the pulmonary arteries may be noted secondary to increased LA pressure leading to pulmonary hypertension. Arrhythmias are not uncommon, especially atrial fibrillation. A left MEA shift is common. Echocardiography will show a dilated LA, LV, and pulmonary veins. Systolic dysfunction may occur.

Cor triatriatum is the term for a heart that develops with 3 atria or an abnormal membrane across 1 atrium, restricting flow into the heart.
result from volume overload caused by mitral regurgitation.

**Management**
Therapy is directed at control of left-sided CHF with conventional medical therapy. Valve replacement is the only surgical option.

**Cor Triatriatum**
Cor triatriatum is the term for a heart that develops with 3 atria or an abnormal membrane across 1 atrium, restricting flow into the heart. Cor triatriatum dexter is the term used when the RA is affected and the abnormal septum obstructs venous return from the vena cava (FIGURES 4 AND 5). Increased pressure caused by the obstruction leads to ascites and signs of right-sided CHF.

Cor triatriatum dexter has been reported in dogs, but not in cats. Presentation for diagnosis is related to the degree of obstruction, with mild obstructions remaining undiagnosed for years. The condition can be palliated with balloon dilation of the abnormal membrane or managed with medical therapy if not severe.6

Cor triatriatum sinister, or supravalvular stenosis, refers to abnormal septal formation across the LA or creation of a second LA. This condition, along with supravalvular mitral stenosis, has only been reported in cats (FIGURE 6).7,8 This condition leads to left-heart CHF due to pressure overload of the pulmonary veins. In the case of cor triatriatum sinister, standard cardiac catheterization is difficult because anatomic features prevent passage of a catheter from the arterial circulation retrograde to the LA. A hybrid open-heart surgery and balloon dilation procedure has been reported with some success.9 Most clients elect medical management of CHF based on cost.

**FIGURE 5.** Right lateral projection of fluoroscopic contrast angiogram. The radiographic marker at the top right is 1 cm across the bottom. One 6-French (~2 mm) pigtail catheter is in the caudal vena cave just behind the caudal RA; a second is in the cranial RA. A radiographic contrast solution has been injected into each catheter. Marked pooling of contrast can be seen in the caudal RA, while reasonable dilution in the cranial RA indicates more movement of blood. The main pulmonary artery is labeled MPA.

**FIGURE 6.** Dual long-axis echocardiographic/Doppler echocardiography images of a kitten with cor triatriatum sinister. The patient’s cranial aspect is on the right side; caudal is on the left. Depth in centimeters is along the left edge of the image. The arrows in each image indicate the location of the abnormal septum. The color Doppler image (B) shows high-velocity turbulent blood flow moving from left atrium 1 (1) into left atrium 2 (2). This flow is returning to the LA from the lungs through the pulmonary veins. It is typically low pressure and low velocity; however, the obstruction is creating excess pressure on the pulmonary veins. The left ventricular outflow tract is labeled LVOT.
R-to-L PDAs have been reported in dogs and rarely in cats. They usually go undiagnosed for the early years of life. At presentation, these patients show signs of exercise intolerance, lethargy, syncope, shortness of breath, and potentially sudden death.

**CYANOTIC CONDITIONS**

Congenital cardiac defects that cause abnormal mixing of deoxygenated and oxygenated blood are collectively identified as cyanotic conditions or “right-to-left” (R-to-L) shunts. The “right” side includes the RA, RV, and PA, while the “left” refers to the systemic circulation, including the LA, LV, and aorta. These conditions, including TOF, “reversed” PDA, and R-to-L shunting septal defects, can lead to hypoxemia and resultant polycythemia.

A significant cause of R-to-L shunting is Eisenmenger’s physiology, which describes a situation in which pulmonary arterial hypertension is greater than that of the systemic circulation. In these cases, an existing left-to-right (L-to-R) shunt, as seen with a typical ventricular septal defect (VSD) or PDA, can “reverse” so that blood is shunted right to left. When this occurs in human medicine, a heart-lung transplant is necessary.

Reversed Patent Ductus Arteriosus

The term reversed PDA is used to describe a PDA that shunts blood from the PA to the aorta, adding deoxygenated blood to the systemic circulation of the caudal body. Conventional theories of the pathogenesis center on the patient having a congenital PDA; chronic high-volume flow in the pulmonary arterial tree is thought to induce a loss of compliance, leading to increased resistance in the PA and a change in the shunt to right to left. It is also theorized that in some cases, the PDA does not reverse, but never actually changes from fetal circulation at all because the pulmonary vascular resistance does not fall with the expansion of the lungs. The term for the latter scenario is persistent fetal flow, rather than reversed PDA.

Regardless of the pathogenesis, the result is an R-to-L PDA that shunts deoxygenated blood from the PA into the descending aorta. The brachiocephalic trunk and left subclavian arteries exit the aorta upstream of the PDA and are unaffected by the shunt. Therefore, the partial pressure of arterial oxygen (\(\text{PaO}_2\)) is reduced only in the caudal portion of the patient, except under stress. The measured \(\text{PaO}_2\) is inversely proportional to the volume of the shunt through the PDA; the greater the shunt volume, the lower the \(\text{PaO}_2\). The decreased \(\text{PaO}_2\), in turn, stimulates the renal system to release erythropoietin, leading to polycythemia. Unless another comorbidity exists, the patient will eventually succumb to chronic hypoxia and polycythemia.

**Clinical Signs and Physical Examination Findings**

R-to-L PDAs have been reported in dogs and rarely in cats. They usually go undiagnosed for the early years of life. At presentation, these patients show signs of exercise intolerance, lethargy, syncope, shortness of breath, and potentially sudden death. Murmurs are not typically appreciated with R-to-L shunting PDA. This is partly due to the decreased pressure gradient between the PA and aorta created by pulmonary hypertension and the hyperviscosity of the blood induced by polycythemia. In some cases, a loud S2 sound may be appreciated related to the crisp closing of the pulmonic valve due to pulmonary hypertension. A jugular pulse can occur with significant tricuspid regurgitation. A positive hepatojugular reflux test indicates elevated RA pressure.

Generalized cyanosis may be present during exercise or excitement. The classic physical examination finding is differential cyanosis, in which the cranial mucous membranes are pink but the caudal tissues are cyanotic. Hindlimb weakness is also a common sign in patients with R-to-L shunting PDA.

**Diagnostic Tests**

Diagnostic tests include radiography, complete blood count, serum chemistry, and echocardiography. Radiographically, evidence of RA and RV enlargement and overcirculation of the pulmonary arteries with a prominent pulmonary trunk are seen. Some tortuosity
of the pulmonary arteries may be appreciated. A prominent bulge is also seen in the descending aorta at the level of the PDA. Signs of left-sided CHF are uncommon. The ECG generally indicates RV enlargement patterns with right MEA shifts. Arrhythmias are variable.

Echocardiography is typically diagnostic, revealing RV hypertrophy and dilation, RA dilation, and dilation of the pulmonary trunk. The use of Doppler echocardiography may allow visualization of the shunt flow in the distal PA. Spectral Doppler echocardiography of any tricuspid regurgitation can yield the estimated pulmonary arterial pressure, which is used for prognosis and response to therapy information. Definitive diagnosis is achieved by performing contrast echocardiography over the abdominal aorta (BOX 2 AND FIGURE 7). As the saline is injected into the vein, it travels to the RA, RV, and PA. A portion of the air bubble contrast will pass through the PDA and descend to the abdominal aorta. Cardiac catheterization is not commonly performed but can be used to measure pulmonary pressure. Arterial blood gas of the femoral artery will indicate the severity of the drop in PaO₂.

**Management**

Medical management is the best approach. Surgical closure of the PDA in the presence of pulmonary hypertension typically leads to death from RV pressure overload, often at the time of surgery. However, surgery has been successfully performed in certain situations in both dogs and cats.¹⁰⁻¹² The priority of therapy is control of erythrocytosis and hyperviscosity. Phlebotomy can be helpful in resolving clinical signs associated with hyperviscosity, such as hindlimb weakness and shortness of breath.¹³ The target packed cell volume (PCV) in these patients is 62%. To achieve
this PCV, approximately 10% of the patient’s blood volume is initially removed. No replacement fluids are given. After several hours of cage rest, an additional 5% to 18% of the blood volume is removed based on the initial PCV.13

Administration of the chemotherapy agent hydroxyurea has been successfully used to retard the production of blood components.14 Complete blood counts and platelet counts are initially monitored biweekly, then periodically to maintain a PCV of approximately 60%. Dosage and administration are customized to minimize the side effects of vomiting, anorexia, alopecia, pruritus, and potential bone marrow hypoplasia.

Other therapies such as vasodilators are of little effect due to their predominantly systemic effects. Sildenafil is a notable exception, as it works by increasing pulmonary arterial vasodilation more than systemic. Sildenafil has anecdotally been described as improving clinical signs. With phlebotomy or hydroxyurea therapy, patients may do well for some years, but the long-term prognosis is poor.

**Tetralogy of Fallot**

Tetralogy of Fallot is named for Dr. Étienne-Louis Arthur Fallot, who first linked the condition to cyanotic babies, although it had been previously described in the medical literature.15,16 Affected neonates have a heart with 4 defects, hence the term *tetralogy*. The defects are (1) obstruction of the right ventricular outflow tract (RVOT); (2) a large VSD, which (3) allows the aorta to shift toward the RV (dextrorotation); and (4) RV hypertrophy. This CHD is not actually 4 problems in one patient, as commonly believed.

**Pathophysiology**

In the embryo, the heart begins as a hollow organ that divides into 4 chambers by the intersection of 3 septa: the spiral, which creates the great vessels and is most superior; the conotruncal, which separates the upper portions of the ventricles, contributes to the semilunar valves, and is centrally located; and the muscular, which separates the lower muscular portion of the ventricles. TOF results when the conal septum forms shifted anterior, superior, and rightward and incompletely fuses with the muscular septum.

If the development of the septum is correctly placed, even if a large VSD is present, the flow may be physiologically balanced. In this situation, the patient may exhibit few signs at rest. The most malignant version shows complete pulmonary atresia, and all blood flow to the lungs is achieved by either the ductus arteriosus before it closes or another systemic-pulmonary shunt. In human medicine, these children are identified in utero and surgical repair is planned accordingly. In veterinary medicine, these patients often die suddenly hours to days after birth.

**Clinical Signs and Physical Examination Findings**

The condition is reported in both cats and dogs. Keeshonds are predisposed. Patients often show a failure to thrive in the early weeks of life and usually present for lethargy, exercise intolerance, syncope, or shortness of breath. During the physical examination, cyanosis may be apparent if the PaO₂ is sufficiently low. Some patients with TOF only show cyanosis during exercise. A murmur may or may not be present. As with R-to-L PDA, hyperviscosity may retard the development of turbulence of a murmur. If a murmur is present, it is generally systolic, located over the left heart base, and of variable loudness related to the PS. Murmurs at the right heart base are either radiation of the PS murmur or flow across the VSD. Under stress, patients may gasp and have pulmonary crackles, which are not related to pulmonary edema, but their genesis is not understood.

Diagnosis typically centers on radiography and echocardiography. An ECG typically shows a right MEA axis shift in sinus rhythm, but MEA may be normal. If the disease is advanced, atrial fibrillation or other arrhythmias may be noted. Ventricular arrhythmias may be present if hypoxemia is severe. Radiographs demonstrate undercirculation of the lungs,
diminished LA size, and normal cardiac size, with possible rounding of the RV. The main PA is small and no poststenotic dilation occurs as with lone PS. The echocardiogram will allow visualization of the VSD, evidence of RV hypertrophy, and malformed RVOT (Figure 8). The LA and LV size will be related to the amount of pulmonary circulation through the RVOT. If the RVOT is very narrow, the LA and LV will be diminished. A Doppler study shows turbulent flow exiting the RVOT and flow across the VSD into the aorta. A contrast echocardiogram can be used to verify R-to-L shunting.

Management

Treatment of TOF involves corrective surgery and is commonly done with great success in human medicine; Olympic snowboarder Shaun White is a notable example. Surgical repair is achieved by closing the VSD and opening the RVOT via cardiopulmonary bypass. Palliative surgery using the Blalock-Taussig procedure, which uses the subclavian artery to create a systemic to PA shunt, has been performed in dogs and cats.17,18 Medical management is used with any intervention. The same approach as with R-to-L PDA of phlebotomy or control RBC production are the priorities. The same approach as with R-to-L PDA of phlebotomy or control RBC production are the priorities. The long-term outcome is directly related to the amount of pulmonary blood flow and ranges from failure to thrive (pulmonary atresia with compensating shunt) to exercise intolerance (mild to moderate RVOT flow).

CHF is uncommon; rather, most patients without corrective surgery succumb to chronic hypoxia, chronic hyperviscosity, cardiac arrhythmias, and sudden death.

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References

CONTINUING EDUCATION

A Look at Unusual Congenital Heart Defects in Dogs and Cats

LEARNING OBJECTIVES
After reading this article, the well-versed veterinary nurse will be able to describe how a right-to-left shunt causes hypoxemia and which right-to-left shunts might be identified in the clinic. The reader should also be able to define several uncommon but important congenital heart defects. The astute nurse will explain the physiology of the 2 groups of defects presented.

TOPIC OVERVIEW
This article is an overview of 6 uncommon congenital heart defects and the associated physiology changes of the cardiovascular system. Summaries of the clinical presentations and management are included.

1. The overall incidence of all congenital heart disease in dogs is approximately:
   a. <1%
   b. 5%
   c. 7%
   d. 10%

2. Which of the following influences stroke volume?
   a. Heart rate
   b. PaO₂
   c. Cardiac output
   d. Preload

3. Eisenmenger’s physiology describes which of the following?
   a. The decrease of heart rate after closure of a patent ductus arteriosus (PDA).
   b. The genetic defect responsible for reversed PDA and tetralogy of Fallot.
   c. The decrease in stroke volume associated with a ventricular premature complex and the sudden increase afterward.
   d. The increase in pulmonary arterial pressure that causes blood to shunt from right to left with reversed PDA.

4. Tricuspid valve dysplasia is most common in which of the following breeds?
   a. Golden retriever
   b. Labrador retriever
   c. Boxer
   d. Greyhound

5. Which of the following is a physical examination finding for a right-to-left shunting patent ductus arteriosus?
   a. Pale mucous membranes
   b. Left basilar grade 5/6 continuous murmur
   c. Differential cyanosis
   d. Hyperkinetic pulses

6. Which of the following chambers is affected in a patient with cor triatriatum dexter?
   a. Left atrium
   b. Right atrium
   c. Left ventricle
   d. Right ventricle

7. Pulmonic stenosis and a large ventricular septal defect are noted in a patient during echocardiography. The ultrasonographer says the aorta is almost hanging in space, and the right ventricle is gigantic. Which defect is most likely?
   a. Mitral valve dysplasia
   b. Eisenmenger’s syndrome
   c. Cor triatriatum sinister
   d. Tetralogy of Fallot

8. Which finding is common in patients with tetralogy of Fallot or a right-to-left shunting patent ductus arteriosus?
   a. Polycythemia
   b. Anemia
   c. Leukocytosis
   d. Thrombocytopenia

9. Tetralogy of Fallot may be palliated with which of the following procedures?
   a. Jackson-Henderson technique
   b. Simple surgical ligation
   c. Blalock-Taussig technique
   d. There are no palliative procedures, and a heart transplant must be done.

10. A right-to-left shunting defect of any kind can cause cyanosis directly related to which of the following?
    a. Increased partial pressure of arterial carbon dioxide
    b. Decreased partial pressure of arterial carbon dioxide
    c. Increased partial pressure of arterial oxygen
    d. Decreased partial pressure of arterial oxygen