CONGENITAL HEART DISEASE
Revised by V'19 cardio group from Dr. John Rush

Introduction

Congenital heart disease can be defined as any cardiovascular malformation present at birth, typically resulting from genetic/hereditary factors and/or environmental causes. It is estimated that 5-10 dogs per 1000 births have congenital heart disease.

An understanding of cardiac embryologic development is very useful in understanding the causes of these defects and resulting pathophysiology. As a review, the embryo's pulmonary circulation is a high resistance system, while the placenta has low resistance. Cardiac output is 2-4X higher than that of an adult and since the developing lungs are not yet equipped to oxygenate the blood, the umbilical vein serves as the embryonic source of oxygen. Several structures exist within the developing cardiovascular system that should not remain permanent (e.g. Foramen ovale, ductus arteriosus, umbilical arteries, umbilical vein, ductus venous).

An accurate diagnosis is important in these animals who are typically young, and often asymptomatic at the time of their first physical exam. Detection of a congenital heart disease could have serious implications for the owner if a purchase return is warranted, breeding potential of the animal is affected, surgical correction is needed, or prognosis is poor.

Detection of a Congenital Heart Defect:

- **Murmurs:** Loud murmurs of long duration are usually the result of cardiac malformation and should be investigated. Characterization of murmurs with respect to timing, location on the chest wall, intensity, quality, radiation, and presence or absence of abnormalities in transient sounds (S1, S2, S3, or S4), is important in differentiation of congenital heart defects.
    - Innocent murmurs in small animals are typically systolic, of short duration, and low intensity (grade III or less). These murmurs frequently change in character or disappear completely with changes in the animal's body position (i.e., lie them on their back and auscultate). Innocent murmurs become softer as the animal ages and usually disappear by 12 months of age.
- The physical examination should also include:
    - Palpation of the precordium to find the site of the apex beat and for thrills
    - Palpation of femoral arterial pulse quality
    - Observation of the jugular veins for distention or pulsation
    - Abdominal palpation for hepatomegaly or ascites
    - Assessment of mucous membrane color and capillary refill time
- The cardiovascular work-up for congenital heart disease may include:
    - Echocardiography is often the first step after physical exam
    - Thoracic radiographs or electrocardiography
    - Blood oxygen content (oximetry) - allows identification of left-to-right or right-to-left shunting defects and provides a quantitative assessment of the degree of shunting.

- Non-selective or selective angiography- Indicated to document cardiac defects and localize the lesion.
- Selective cardiac catheterization to obtain direct measurement of intracardiac pressures and blood oximetry

I. <u>Patent Ductus Arteriosus (left to right shunting)</u>- dogs, cats, horses, llamas

Description:

In the fetus, blood returning to the heart either passes through the foramen ovale to the left atrium or is pumped through the right ventricle to the pulmonary artery. The pressure in the pulmonary vessels is high during fetal life, so blood pumped into the pulmonary artery is shunted through the ductus arteriosus to the aorta, preventing unneeded pulmonary circulation. At birth, expansion of the lungs causes a drop in pulmonary vascular resistance and the pressure in the systemic circuit rises. This results in a reversal of blood flow through the ductus arteriosus (blood now flows from the aorta to the pulmonary artery). This reversal, coupled with the inhibition of prostaglandin synthesis, and local increase in PO2 results in vasoconstriction of the ductus with functional closure by 72 hours postpartum in dogs and cats. Failure of ductal closure results in patency (patent ductus arteriosus - PDA) and a permanent connection from the aorta to the pulmonary artery.

Signalment

- PDA is most common in female purebred dogs
- A polygenic mode of inheritance has been proven in the poodle
- Predisposed dog breeds: poodles, Pomeranians, collies, chihuahuas, Maltese, Shetland sheepdogs, German shepherds, cocker spaniels, and Irish setters
- Siamese cats may also be predisposed to PDA.
- May present for evaluation of poor growth, exercise intolerance, or respiratory distress. 25-35% of dogs will present with signs of congestive heart failure.

Clinical Findings

- Continuous cardiac murmur at the left heart base, which peaks in intensity at the second heart sound.
  - The systolic component of the murmur radiates and may be auscultated over the entire thorax.
- Presence of a thrill (especially left heart base)
- Bounding arterial pulses
- Left sided cardiac enlargement and eccentric hypertrophy
  - Mitral valve regurgitation, and the accompanying systolic, regurgitant quality murmur, frequently develops secondary to left ventricular dilation and stretching of the mitral valve annulus.
- Increased filling pressures in the LA and LV, which may progress to pulmonary edema.

Diagnosis

- **Thoracic radiographs**: marked left sided cardiomegaly, enlarged pulmonary vasculature (pulmonary overcirculation), dilation of the descending aorta, dilation of the main pulmonary artery, left auricular enlargement.

- If the patient has progressed to heart failure an interstitial or alveolar lung pattern may be visible.

- **ECG**: Left ventricular enlargement pattern and p-mitrale, sinus tachycardia, ventricular or supraventricular arrhythmias, atrial fibrillation.
- **Echocardiogram:** Left atrial and ventricular enlargement, prominent aortic root motion, dilation of the pulmonary artery.
    - Using color flow doppler, the PDA may be visualized as continuous flow in the main pulmonary artery
- **Treatment:**
    - Ductal occlusion or surgical ligation of the ductus, preferably before 4-6 months of age, is the treatment of choice, and is usually curative in young animals who have not yet developed CHF.
    - Mitral insufficiency may resolve with improvement in cardiac dilation after ductal occlusion, and as young animals "grow into" their mitral valve.
    - Animals with arrhythmias, long-standing congestive heart failure prior to surgery, or surgical correction after 1 year of age often require long-term medical therapy.
    - Prognosis for uncorrected PDA is poor- 40-70% mortality rate by one year.

II. <u>Pulmonic Stenosis (PS)</u> - dogs

Description:

Pulmonic stenosis is usually due to valvular stenosis/dysplasia or subvalvular stenosis. In dogs, valvular stenosis is the most common cause, and can be recognized as thickened, malformed valves. Subvalvular stenosis may accompany valvular stenosis or occur as an isolated lesion, and is characterized by a thick fibrous ridge of tissue below the pulmonic valve in the RV outflow tract.

Narrowing of the pulmonic valve obstructs ejection of blood from the RV. In response, the RV undergoes hypertrophy to maintain cardiac output across the narrowed pulmonary outflow tract (pressure overload = concentric hypertrophy). Secondary RA enlargement may develop.

Signs may result from forward heart failure (syncope, muscular weakness) or backward right sided heart failure (ascites, jugular venous distension/pulsation). Forward heart failure occurs when the RV is unable to pump a sufficient volume of blood through the lungs to the LV. The result is a decrease in left ventricular preload and an inability to increase CO in response to exercise/excitement. RV myocardial failure, tricuspid regurgitation, or pulmonic valve insufficiency may contribute to development of right sided heart failure.

Signalment

- Predisposed dog breeds: English bulldog, Schnauzer, Beagle, Chihuahua, Terriers, Cocker Spaniel, Samoyeds.
- Often 3-7 years old at the onset of clinical signs

Clinical Findings

- Most animals are asymptomatic. The presence of a heart murmur leads to cardiac workup.

- Murmur- Loud, harsh, ejection quality murmur, with point of maximal intensity at the left heart base
    - A second regurgitant quality murmur may be auscultated on right hemithorax if tricuspid insufficiency has developed
- Presence of a thrill
- Mucous membrane and pulse quality can range from normal in cases of mild stenosis, to pale with weak pulses in critical cases.
- Hepatomegaly, ascites, pleural effusion, and jugular venous distension are possible

Diagnosis:

There are many useful diagnostic tools for identifying pulmonic stenosis, however it is important to note that the severity of the defect cannot be determined without cardiac catheterization or Doppler echocardiography.

- Grading system (in dogs):
    - Mild: 10-50 mmHg - typically asymptomatic and do not require therapy, but intervention may be considered if clinical signs are present
    - Moderate: 50-100 mmHg - candidates for balloon valvuloplasty or surgery if clinical signs or concurrent defects (ASD, tricuspid dysplasia, etc.)
    - Severe: >100 mmHg - candidates for balloon valvuloplasty or surgery; if clinical signs are present, these dogs do not typically live beyond 4-7 years without treatment.
- **Thoracic radiographs:** Right ventricular enlargement and dilation of the main pulmonary artery on VD radiograph. Right atrial enlargement is variably noted. If the patient has progressed to right heart failure, caudal vena cava enlargement may also be noted.
- **ECG:** Often normal in cases of mild stenosis. Moderate-severe pulmonic stenosis can cause a right ventricular enlargement pattern and/or right axis shift, while P-pulmonale (right atrial enlargement pattern) is less frequently recorded. Ventricular or supraventricular arrhythmias may also occur.
- **Echocardiogram:** Right ventricular hypertrophy, hypertrophied interventricular septum, narrow pulmonary outflow tract, thickened pulmonic valve leaflets, post-stenotic pulmonary artery dilation,
    - Paradoxical septal motion, wherein the interventricular septum moves towards the RV during systole, may be noted on echocardiogram.
    - Doppler echo can be used to document turbulent blood flow in the main pulmonary artery and in many cases will provide an accurate estimate of the pressure gradient across the pulmonic valve.
- **Angiography:** Post stenotic dilation and right ventricular hypertrophy can be documented using both selective and non-selective angiography, but in order to differentiate between valvular and subvalvular stenosis, selective angiography is often required.

Treatment:

- Several procedures have been developed to enlarge the pulmonic valve orifice. The appropriate technique for each patient is selected based on location and type of stenosis

- Balloon valvuloplasty is associated with lower mortality than surgery, making it the first intervention of choice in most cases.
- Surgical procedures: Bistoury and valvotomy techniques, patch graft techniques

- Medical management is restricted to antiarrhythmic medications (including beta blockers to reduce outflow obstruction), and controlling signs of congestive heart failure using diuretics and a low-salt diet.

III. <u>Aortic Stenosis (AS)</u> - dogs, cats, pigs

Description: Aortic stenosis can result from narrowing at the subvalvular, valvular, or rarely supravalvular aortic outflow tract. Subvalvular aortic stenosis (SAS) is by far the most common type in dogs, and is characterized by a thick band of fibrous connective tissue just below the aortic valve. Valvular stenosis results from dysplastic valves with thickened or fused leaflets.

Because aortic stenosis causes obstruction to ventricular outflow, the left ventricle must hypertrophy to maintain normal stroke volume (pressure overload = concentric hypertrophy). Turbulent blood flow, generated by the stenosis, causes a murmur and post-stenotic dilation distal to the site of obstruction. Advanced disease and left ventricular failure leads to left atrial enlargement and eventually pulmonary edema (more likely in cats, than dogs). Mitral or aortic insufficiency may accompany and complicate aortic stenosis.

Sequelae of AS include syncope/sudden death, aortic endocarditis, and less commonly left sided CHF.

Signalment:

- Predisposed dog breeds: Newfoundland, GSD, Boxer, Golden Retriever, Rottweiler, and German Shorthaired Pointer
  - In the Newfoundland, aortic stenosis is transmitted genetically.
- Many dogs with subvalvular stenosis are asymptomatic at the time of presentation, but some may present for syncope, weakness, stunted growth, exercise intolerance, dyspnea or cough.
- Aortic stenosis is less commonly encountered in cats. However, cats with this defect frequently develop heart failure or sudden death before one year of age.

Clinical Findings:

- The first outward clinical sign of subvalvular aortic stenosis in dogs can be syncope or sudden death. These severe signs are the combined result of an inability to increase stroke volume in response to exercise, and compromised myocardial perfusion.
  - SAS limits the ability of the left ventricle to increase stroke volume in response to increased tissue demands, so increases in cardiac output result largely from increases in heart rate. Ventricular hypertrophy, combined with increased heart rate, results in an increased myocardial oxygen demand. Unfortunately, elevated intraventricular pressures limit the coronary arterial blood flow during systole and tachycardia shortens diastole, therefore myocardial oxygen demand increases while coronary artery perfusion decreases leading to left ventricular endocardial ischemia. Myocardial ischemia leads to myocardial depression, decreased myocardial compliance, myocardial necrosis and fibrosis, and increased risk for arrhythmia.

- Murmur- Systolic, ejection quality murmur, with point of maximal intensity at the left heart base (3rd-4th intercostal space). The murmur may be equally loud at the right heart base and may radiate up the carotid arteries.
- Presence of a thrill
- Prominent left apical impulse on palpation
- Weak, slow-rising femoral pulses (pulsus parvus et tardus)
- +/- Arrhythmias and pulse deficits

Diagnosis:

- **Thoracic radiographs:** Left ventricular enlargement, dilation of the aorta, +/- left atrial enlargement and evidence of congestive heart failure in advanced cases
- **ECG:** Normal in most cases. When abnormal results are found, findings vary with severity of stenosis.
    - Increased QRS amplitude and/or duration may be present, indicating LV enlargement, while ST segment depression suggests myocardial ischemia, necrosis or fibrosis. After exercise, ST segment depression or ventricular arrhythmias indicate exertional myocardial ischemia.
- **Echocardiogram:** Echo-dense band/ridge of tissue in LV outflow tract in dogs with subvalvular aortic stenosis. Abnormal valvular anatomy or motion with valvular stenosis. Concentric LV hypertrophy with prominent papillary muscles. Post stenotic dilation.
    - Doppler echo documents turbulent blood flow in the ascending aorta and can be used to measure the pressure gradient across the aortic valve:
        - Mild AS = 10-40 mmHg
            - Typically asymptomatic; no therapy required
        - Moderate AS = 40-80 mmHg
            - May live a normal lifespan and/or die suddenly
        - Severe AS = >80 mmHg
            - Many dogs will develop clinical signs or shorten lifespan
- **Angiography:** Selective angiography documents valvular and subvalvular anatomy, LV hypertrophy, post-stenotic dilation and presence/absence of aortic insufficiency.

Treatment:

- Surgical treatment: Surgical correction of subaortic stenosis is rarely attempted because the technique requires cardiopulmonary bypass and is associated with a high surgical risk and may not dramatically prolong survival time. Animals who are considered surgical candidates are those with remarkable clinical signs, severe arrhythmias, ECG evidence of ischemia, and gradients in excess of 80 mmHg, who are thought to be at risk of sudden death
    - Balloon valvuloplasty is sometime performed in dogs with SAS, but the balloon less effective in consistently stretching the fibrous tissue ringas durable as what is seen in animals with pulmonic stenosis. This technique is most useful in animals with valvular aortic stenosis and those with a discrete subvalular ring. A cutting balloon might improve the effectiveness of balloon valuloplasty.
- Medical management:
    - Atenolol or other beta-adrenergic blocking drugs can be used in an attempt to reduce the myocardial oxygen demand and diminish the risk of arrhythmic death.

These drugs are often used in dogs with severe stenosis or ventricular arrhythmias but their efficacy has not yet been supported by the literature, and 1 study failed to show improvement.

- Prophylactic antibiotic use is indicated for any procedure associated with bacteremia (e.g. dental cleaning), as aortic stenosis can predispose patients to aortic valve endocarditis.

IV. <u>Ventricular Septal Defect -</u> cats, dogs, horses, cattle, alpaca, small ruminants, pocket pets

Description:

Malformation of the interventricular septum can result in ventricular septal defects that allow communication between left and right ventricles. While a VSD can occur anywhere in the septum, it is usually located high in the membranous portion of the septum (below the aortic valve). This defect may occur alone or as part of another defect (e.g. tetralogy of Fallot).

VSDs tend to be left-to-right shunting because of the higher systolic pressure in the left ventricle. In these typical cases of high, left-to-right shunting VSDs, blood is ejected from the LV into the right ventricular outflow tract causing an increase in blood volume moving through the pulmonary circulation and into the LA. This volume overload will cause the left ventricle to undergo eccentric hypertrophy. In some cases, the missing septal tissue contains supporting structures for the aortic valve; aortic insufficiency can result, contributing additional volume overload to the LV.

Signalment

- Predispositions: VSD is the most common congenital heart defect in horses, cattle and sheep and is also among the most common in cats. In dogs, only the keeshond and English bulldog are predisposed.
- Microphthalmia may be seen in association with VSD in both dogs and calves.
- Dogs with large VSDs and heart failure tend to develop pulmonary edema while cats are more prone to biventricular failure which leads to both pulmonary edema and pleural effusion.
- Small to medium sized VSD is often well tolerated in dogs and cats.

Clinical Findings

- Clinical signs are generally observed in animals with large ventricular septal defects, as patients with small VSDs tend to be asymptomatic
    - Weakness, cough and respiratory distress
    - Murmur- harsh, holosystolic, loudest at the right cranial sternal border
    - Arterial pulses are usually normal unless the animal has entered CHF
    - Jugular venous distention may be present if the VSD is complicated by pulmonary hypertension or biventricular CHF.
    - Small VSDs may spontaneously close in the first 2 years of life, while animals with large defects may develop CHF before they are 1.5-2 years old.

Diagnosis

- **Thoracic radiographs**: Often normal, or may demonstrate left heart enlargement, pulmonary edema, and variable right ventricular enlargement.

- **ECG:** Normal with small VSDs. Larger defects may show evidence of LA or LV enlargement, right bundle branch block, or arrhythmias.
- **Echocardiogram:** Left atrial enlargement, hyperdynamic, dilated left ventricle. If the VSD is large enough it can be visualized as an echo lucent region at the top of the ventricular septum.
    - Doppler echo can be utilized to easily identify blood flow through the defect and to confirm the direction of blood flow.
    - Pulmonary:systemic blood flow can be estimated.
- **Selective cardiac catheterization**: The diagnosis can be confirmed, the direction of shunting demonstrated, and the functional integrity of the aortic valve established using this method. Oximetry data, obtained during catheterization can be used to calculate the magnitude of the shunt. The size of the shunt can be expressed as the ratio of the blood flowing through the pulmonary and systemic circulation. If pulmonary:systemic blood flow ratio is > 2.5, the patient is at risk of developing pulmonary vascular disease or heart failure, and surgery is recommended.

Treatment:

- Surgical treatment: Surgery should be considered if clinical signs are present or if the defect is very large (large volume shunted blood).
    - Surgical closure of a VSD requires open heart surgery which is not feasible at many institutions.
    - Pulmonary artery banding is a palliative technique wherein the pulmonary vascular resistance is increased to decreased the magnitude of the left-to-right shunt. However it is important to note that if the band is too tight, right-to-left shunting can occur.
    - Surgery is likely not required for animals who are asymptomatic, have minimal cardiomegaly, no evidence of CHF, or have pulmonary:systemic blood flow ratio < 2.
    - If the VSD is lower in the interventricular septum then a catheter-based closure technique may be feasible.
- Medical management may be suitable for patients with signs of CHF
- Prevention: Affected and related animals should not be bred.

V. <u>Atrial Septal Defect (ASD)</u> - cats, dogs, horses, cattle, llama, alpaca

Description:

Malformation of the atrial septum allows abnormal blood flow between the two atria. These defects are described as ostium primum or ostium secundum; ostium primum describes a failure of atrial tissue formation low in the atrial septum, and ostium secundom defects are located higher, in the middle of the atrial septum. In cats and dogs, atrial septal defects rarely occur alone. Instead they tend to be seen in association with other congenital heart defect such as endocardial cushion defect, pulmonic stenosis, or tricuspid dysplasia.

The pathophysiology of ostium primum and ostium secundum defects are similar: blood is typically shunted from the left atrium to the right atrium through the defect, increasing the volume of blood being pumped through the right heart, the pulmonary vasculature and into the left atrium. The clinical magnitude of the shunt is determined by the size of the defect and the

diastolic compliance of the ventricles, so that small atrial septal defects tend to be asymptomatic while large defects result in right atrial and ventricular dilation due to volume overload. If pulmonic stenosis, tricuspid dysplasia or right ventricular failure is present concurrently, pressure in the RA may rise enough to cause reverse (right-to-left) shunting through the defect.

Signalment

- Predisposed dog breeds: boxer, Samoyed, Doberman pinscher, old English sheep dog
- May present for exercise intolerance, respiratory distress, syncope or lower respiratory tract infections

Clinical Findings

- No murmur is generated by blood flow through an ASD, however auscultatory abnormalities may be present due to the increased volume of blood passing through the right heart:
    - Systolic murmur of relative pulmonic stenosis
    - Low intensity, rumbling diastolic murmur of relative tricuspid stenosis
    - Fixed splitting of the second heart sound.
- Most dogs with ASD remain asymptomatic and, due to the lack of a loud murmur, some probably go unrecognized.

Diagnosis

- **Thoracic radiographs:** Usually normal with small ASDs. Large ASDs may cause visible right heart enlargement, pulmonary vasculature enlargement, and variable degrees of LA enlargement.
- **Electrocardiogram:** Typically normal. Right ventricular enlargement pattern may occur in large shunts.
- **Echocardiogram:** The most useful non-invasive diagnostic technique for ASD. Large ASDs can be visualized as an echo lucent space in the atrial septum.
    - Contrast (e.g. agitated saline) or Doppler echocardiography can be useful in documenting right-to-left shunting or bidirectional blood flow across the atrial septum.
- **Cardiac catheterization:** Documents the presence, direction, and magnitude of shunt flow
- **Angiography:** selective angiographic studies from pulmonary arterial injections or after passage of the catheter through the defect will also demonstrate the ASD and the direction of blood flow.

Treatment

- Surgical treatment: Usually requires cardiopulmonary bypass techniques. A catheter based- technique is also available to close an ASD using an Amplatz ASD device. When other concurrent defects are present the clinical course will be determined by the combined effects of the defects.
- Medical management: Animals with very large defects may develop heart failure within the first few years of life and can be managed with medical therapy.

VI. <u>Atrioventricular Valve Malformations</u> - cats, dogs, horses

Description:

Malformations of the mitral or tricuspid valves can result in insufficiency, stenosis or both, with valvular insufficiency being most common. In addition to thickened and fused valve leaflets, abnormal chordae tendinae and/or abnormal papillary muscles may be noted. Atrioventricular valve stenosis causes impaired ventricular filling, which may elevate the atrial pressures and lead to congestive heart failure.

AV valve insufficiency results in volume overload in the affected ventricle, leading to eccentric hypertrophy. In cases of mitral valve malformation, both the LV and LA become enlarged, and left-sided CHF (pulmonary edema) may result. Tricuspid dysplasia causes right ventricular hypertrophy (eccentric), and large increases in RA size, which can lead to right-sided CHF (ascites and/or pleural effusion).

Signalment:

- Predispositions:
    - Mitral and tricuspid valve dysplasia is the most common congenital heart defect in cats.
        - Some cats are severely affected at a young age. Others live many years with no evidence of dysfunction.
    - Large breed dogs are also predisposed e.g. Great Dane, German shepherd, Labrador retriever and Weimaraner
- Dogs with AV valve dysplasia tend to present at a young age, with signs of weight loss, exercise intolerance, dyspnea or coughing

Clinical Findings:

- Murmur- pansystolic. Occurs over the affected valve.
    - Mitral dysplasia creates left apical regurgitant quality murmurs that radiate dorsally
    - Tricuspid dysplasia creates a murmur that is loudest on the right hemithorax over intercostal spaces 3-5
    - AV valve stenosis may result in a soft diastolic murmur (left apical for mitral and right apical for tricuspid)
- Dyspnea, tachypnea, pulmonary crackles are present in animals that have developed CHF.
- Jugular venous distension, hepatomegaly, ascites or pleural effusion (signs of right-sided CHF) may be seen in cases of tricuspid valve malformation, while pulmonary edema (left-sided CHF) is possible in cases of mitral valve malformation.

Diagnosis:

- **Thoracic radiographs:** Mitral insufficiency can cause LA and LV enlargement, accompanied by pulmonary venous distension and pulmonary edema when heart failure occurs. With tricuspid dysplasia, RA and caudal vena cava enlargement and pleural effusion may be seen
- **ECG:** Possible findings include P-mitrale, LV enlargement pattern, and supraventricular arrhythmias in cases of mitral dysplasia. Right ventricular and/or right atrial enlargement patterns may be seen when tricuspid dysplasia is present.

- **Echocardiogram:** Abnormal morphology of valve leaflets, chordae tendinae, papillary muscles. LA and LV enlargement noted in cases of mitral dysplasia. Tricuspid dysplasia leads to visible RA and RV enlargement.
    - Color flow Doppler echocardiography can be used to document AV valvular regurgitation
- **Cardiac catheterization:** Tricuspid dysplasia will lead to normal oximetry, elevated RA pressure, and tricuspid regurgitation.

Treatment: AV valve malformation is usually managed medically, but in selected cases, mitral or tricuspid valve replacement can be performed

- Surgical treatment: AV valve replacement requires the use of cardiopulmonary bypass. Unless surgery is performed, if the disease is severe then this progressive disease is typically lethal within a few years.
- Medical management:
    - Arterial vasodilators - reduce mitral regurgitation and increase forward blood flow.
    - Treatment of CHF
    - Digitalis- utilized in animals who are refractory to diuretics and vasodilators, who have diminished myocardial function (as seen on echo), or who have atrial tachyarrhythmias. Other antiarrhythmics may be used as needed.

RIGHT-TO-LEFT SHUNTING LESIONS

I. <u>Tetralogy Of Fallot</u> - cats, dogs, horses

Description

Tetralogy of Fallot (TF) is defined as a cardiac defect with four pathologic findings: 1) pulmonic stenosis, 2) high ventricular septal defect, 3) dextroposition of the aorta such that it overrides the interventricular septum, and 4) secondary right ventricular hypertrophy.

The obstruction to pulmonary outflow forces the right ventricle to develop increased systolic pressures. Blood typically shunts from right-to-left at the level of the ventricular septal defect and in addition, the overriding aorta receives blood from both ventricles. Resultant arterial oxygen desaturation leads to cyanosis and increases in red blood cell mass. Arrhythmias, hypoxemia and complications of polycythemia can cause clinical signs. Systemic embolism may result because thrombi are able to cross the VSD and enter systemic circulation. Congestive heart failure is uncommon.

Signalment

- Predisposed dog breeds: keeshond (genetic), English bulldog
- Most animals are cyanotic at the time of presentation
- May present for exercise intolerance, dyspnea, syncope or stunted growth.

Clinical Findings

- Murmur- any murmurs present may be attenuated in animals w polycythemia, severe pulmonic stenosis, or minimal flow through the VSD.
    - Murmur caused by pulmonic stenosis are heard best at the left base (systolic ejection quality).
    - Ventricular septal defect murmur is loudest at the right cranial sternal border
- Neurologic abnormalities resulting from polycythemia (hematocrit may rise above 75%)

- Hypoxemia can be seen via pulse oximetry or blood gas analysis

Diagnostics

- **Thoracic radiographs:** RV hypertrophy is possible, but some animals with tetralogy of Fallot will have normal radiographic cardiac size. In some animals, decreased vascular markings and pulmonary artery dilation may be observed, while in others, the PA remains small.
- **ECG:** Right ventricular enlargement pattern may be evident
- **Echocardiogram:** Findings usually show high VSD, right ventricular hypertrophy, pulmonic valve/outflow tract abnormalities, and right-to-left shunting with contrast echocardiography or microbubble injection. Dextroposition of the aorta can be visualized in most cases.
- **Cardiac catheterization**: required in some animals to rule Eisenmenger's syndrome and other cardiovascular malformations. Definitely indicated if surgical therapy is contemplated.

Treatment

- Surgical treatment: Indicated in cyanotic animals with clinical signs, most of whom will die by 2-5 years old without surgical intervention. Complete surgical correction is difficult and requires cardiopulmonary bypass, but several palliative procedures exist, which involve anastomosis of a systemic artery to the pulmonary artery (Pott's or Blalock-Tuassig procedure) to increase pulmonary blood flow. Such palliative surgeries can keep the animal comfortable for several years.
- Balloon valvuloplasty of the PS may be considered
- Medical management: Can be used in non-cyanotic animals who have mild pulmonic stenosis. Antithrombotic therapy, periodic phlebotomy, hydroxyurea, and non-selective beta-blocking drugs to manage the polycythemia and decrease right to left shunting.

II. <u>Right-to-left VSD with pulmonary hypertension (aka Eisenmenger's syndrome)</u>– dogs

Description

If a typical left-to-right shunting VSD is very large, ventricular systolic pressures tend to equilibrate due to chronic pulmonary over-circulation leading to severe pulmonary hypertension. If the RV pressure equals or exceeds that of the LV, bi-directional flow or right-to-left shunting will occur, resulting in systemic arterial oxygen desaturation, cyanosis and polycythemia.

Clinical Findings

- Cyanosis (may be exacerbated with exercise), exercise intolerance, weakness, syncope
    - Neurological abnormalities may also be noted as a result of severe polycythemia, bacterial emboli or venous thrombi.
- Murmur – often present but may be soft or absent if polycythemia
- Second heart sound may be loud or split

Diagnostics

- **Thoracic radiographs:** Enlarged right ventricle & enlarged, tortuous pulmonary arteries
- **ECG:** Right ventricular enlargement pattern is usually present
- **Echocardiogram:** contrast echocardiography (bubble study) can be performed to demonstrate right-to-left shunting.

Treatment: Surgical intervention is not indicated and long-term survival, even with medical management, carries a poor prognosis.

- Medical management: Exercise restriction, periodic phlebotomy, and antithrombotic drugs (aspirin or clopidogrel) can be used to reduce the risk of thromboembolism. Sildenafil has also been used to reduce pulmonary hypertension and limit right-to-left shunting.

III. PDA with pulmonary hypertension (Right-to-Left PDA) – dogs

Description:

If the high pulmonary vascular resistance of the fetus fails to regress postpartum, or if pulmonary arteries develop muscular hypertrophy in response to high pressure blood flow through the pulmonary vasculature, pulmonary hypertension will result and lead to bidirectional or right-to-left shunting through the ductus.

Clinical Findings: it is possible for an animal with right-to-left PDA to be asymptomatic for several months to a few years, prior to developing clinical signs.

- Differential cyanosis- Oxygenated blood in the aorta supplies the head resulting in normal, pink mucous membranes. Distal to the site of the ductus, oxygen desaturated venous blood enters arterial circulation, such that abnormal cyanotic mucous membranes can be seen in the caudal portion of the body.
- Weakness, dyspnea, arterial thromboembolism, and right sided heart failure may result.
- Murmur: overall, the murmur is attenuated, and the diastolic component is completely absent. A loud second heart sound may be heart at the left base.

Diagnostics:

- **Thoracic radiographs:** Biventricular enlargement may be noted, and the triad of bumps typical of a left-to-right PDA may be replaced by an aneurysmal dilation in the descending aorta and dilated or tortuous pulmonary arteries. The lung fields appear hypovascular.
- **ECG:** Right ventricular enlargement pattern
- **Echocardiogram**: Right ventricular hypertrophy, PA dilation. Visualization of the ductus can be more difficult due to lower velocity flow. A contrast study of the abdominal aorta can be performed to demonstrate right to left shunting.

Treatment: Surgical intervention is contraindicated because the open ductus provides a “pop-off valve”- ligation of the ductus can fatally increase pulmonary arterial pressure.

- Medical Management: Sildenafil has been used to reduce pulmonary hypertension and limit right-to-left shunting, however these dogs typically have a shortened life-span.

IV. ASD (Right-to-left ASD)

Description

ASD in combination with pulmonic stenosis, tricuspid dysplasia, right ventricular failure, or pulmonary hypertension may produce elevation of the right atrial pressure and lead to reversal of the direction of the shunt. As deoxygenated blood flows from the right atrium to the left atrium, systemic arterial oxygen desaturation results. In contrast to VSD and PDA, atrial

septal defects rarely results in obstructive pulmonary vascular disease and pulmonary hypertension in dogs and cats. When ASD is present in combination with conditions that produce right-to-left shunting, cyanosis, weakness, and arterial thromboembolism may be observed.

ADDITIONAL CARDIOVASCULAR DEFECTS

- **Complex congenital defects:** There are many complex congenital defects wherein, several of the aforementioned defects may be combined, or more severe defects such as failure of development of cardiac chambers may occur. It is likely that many complex congenital cardiac defects in animals are lethal within the first few hours of life and go unrecognized.
- **Vascular ring anomalies:** Vascular ring anomalies result from abnormal embryologic development of the aortic arches. Persistent right fourth aortic arch is the most common in the dog, where German shepherds and Irish setters are predisposed. The esophagus becomes trapped by the base of the heart, the abnormally formed aorta (from the right 4th arch instead of the left), the pulmonary artery, and the ductus arteriosus. Associated clinical signs include regurgitation as puppies start to eat solid food. Surgery is indicated and may be curative if the esophagus is not markedly enlarged at the time of surgery.
- **Peritoneopericardial diaphragmatic hernias:** Reported in dogs and cats, this type of hernia occurs when abdominal organs herniate into the pericardial sac. Animals can present for gastrointestinal disturbances, collapse, or respiratory signs. Surgical therapy is usually curative.
- Additional infrequently observed defects include coarctation of the aorta and ectopia cordis (calf).