Mitral Valve Disease and Cavalier King Charles Spaniels

Degenerative mitral valve disease (MVD) is the leading cause of death of Cavaliers. Other names for the condition include:

- cardiac valve disease (CVD)
- endocardiosis
- atrioventricular valve endocardiosis
- chronic degenerative valvular disease
- chronic valvular disease
- chronic mitral valve insufficiency
- myxomatous atrioventricular degeneration
- chronic valvular fibrosis
- acquired mitral regurgitation or insufficiency,
- mitral valve defect

Veterinary cardiologists began compiling statistics on Cavaliers with MVD murmurs in the United Kingdom in 1990. Since then, cardiologists have examined the hearts of many thousands of Cavalier King Charles, and have found that the percentage of Cavaliers who develop MVD murmurs increases at a rate of about 10% per year. So, roughly 10% of Cavaliers by age one year have MVD murmurs, and 20% aged between one and two years have murmurs, and so on for each age level. More than half of all Cavaliers aged five years have murmurs, and it is the very rare Cavalier at age ten years which does not have, at the very least, a low grade MVD murmur.

What It Is

Mitral valve disease is a uniquely serious, potentially life-shortening problem for Cavalier King Charles spaniels and is their leading cause of death. MVD is the most common heart disorder in older dogs of all breeds, affecting more than a third of all dogs over 10 years of age. Several smaller breeds of dogs are predisposed to MVD. However, in most all breeds, MVD does not result in heart failure, and does not cause death.

In the Cavalier King Charles spaniel, the prevalence of MVD is about 21 times that in the typical breed of dog. Also in Cavaliers, the onset of the disease typically is much earlier in life. It has been reported that, once diagnosed, MVD is much more rapid in Cavaliers than in other breeds, possibly reaching a life-threatening stage within as little as 1 to 3 years, rather than the average 3 to 5 years in other breeds. Studies of Cavaliers have concluded that it has a genetic cause.

MVD is a degeneration and fibrosis of the heart's mitral valve, one of four sets of valves in the heart. The mitral valve is a set of double flaps, called "leaflets", that open and close like a set of one-way doors at appropriate times during each heart beat. Normal mitral valve leaflets are very

thin and nearly transparent. They are connected by tendons (chordae tendineae) to the muscles of the left ventricle in the heart.

As the diseased mitral valve degenerates, it loses its flexibility, its leaflets and struts thicken and shorten, and its fibers stiffen. The leaflets no longer fully close after each pumping action, allowing blood to jet backwards through them from the ventricle (bottom of the heart) back into the atrium (top of the heart). As the condition worsens, more and more blood is able to backflow through the valve, causing both the left atrium and the left ventricle to enlarge. In the final stages, the valve's struts sometimes break, causing the valve to collapse completely.

The increased backflushing of blood causes blood to back up onto the lungs. This backed up fluid can be found in the sac around the heart (the pericardial sac), in the space around the lungs (the pleural space), or even in the lungs themselves (the lungs are soaked with water like a wet sponge). This fluid build-up can cause breathing problems and coughing. Gagging up fluid after coughing can indicate especially serious uncontrolled congestive heart failure.

Diagnosis

The earliest indications of MVD are outwardly invisible and silent and can only be observed by **echocardiography (ultrasound scanning**). The first indication is the excessive bulging of the mitral valve leaflets into the left atrium, which is called mitral valve prolapse (MVP), followed by thickening of the leaflets, and then by the presence of a soft whistling sound, called a "murmur", which can be heard by a veterinarian using a stethoscope, which is called auscultation.

Auscultation. The murmur sound is caused by the turbulent flow of blood jetting backwards through the damaged leaflets of the mitral valve into the left ventricle, increasing pressure in the left atrium. It is loudest on the left side, near the bottom of the chest. Cavaliers should be screened for heart murmurs annually, beginning at age one year (all dogs should have their heart sounds assess at every examination by a veterinarian). Once mitral valve disease is detected, its progression can be monitored with stethoscopic examinations (auscultations), x-rays, echocardiograms, and color Doppler echocardiograms.

X-rays (radiographs). Radiography (x-ray) is used to determine if the heart is enlarged, and if there are signs of congestive heart failure (fluid in or around the lungs). Once MVD is diagnosed, x-rays should be done at least annually and more often if congestive hart failure develops. Chest x-rays are the single test that tells us whether a dog is in congestive heart failure, and one of the most accurate ways to monitor drug therapy.

Heart ultrasound (echocardiography). Echocardiography (ultrasound scanning) used to evaluate heart size, function, and valve appearance. Echo scans can demonstrate the thickened valve leaflets and their abnormal movement, such as prolapse (MVP). The color Doppler can evaluate the direction and velocity of blood flow, quantifying blood leakage. It can be used to distinguish MVD from benign murmurs in ambiguous cases. The Doppler may detect leakage before it is audible as a murmur. However, trivial regurgitation of blood through the mitral valve may be

present in as many as 50% of normal dogs. In such cases, however, there is no MVP or valve thickening present.

Grades of Mitral Valve Disease Murmurs

Mitral valve murmurs are graded from the mildest and least audible, Grade 1, to the loudest and most turbulent, Grade 6. Most Cavalier King Charles spaniels show a gradual progression in the loudness of the MVD murmur.

Grade 1 (I): A Grade 1 murmur can be heard with a stethoscope in a quiet room.

Grade 2 (II): A Grade 2 can be consistently heard with the stethoscope.

Grade 3 (III): Grade 3 murmurs are louder and are heard as soon as the stethoscope is applied.

Grade 4 (IV): Grade 4s are quite loud, and the vibration can be felt with fingertips without a stethoscope.

Grade 5 (V): A Grade 5 murmur is louder, with a precordial "thrill".

Grade 6 (VI): The Grade 6 is so loud it can be heard with the stethoscope removed from the chest, or even without using the stethoscope.

Blood pressure and EKG. While these tests do not diagnose MVD, they should be monitored in dogs with MVD. High or low blood pressure should be treated and monitored if present. Abnormal heart beats as detected by EKG should also be treated and monitored, if present.

Symptoms

The very earliest stages of MVD produce no symptoms. A murmur may be present for months to years prior to leading to symptoms. The first change that occurs, which often goes unnoticed, is increase in the respiratory rate (breaths per minute). IF any dog is taking more than 40 breaths per minute while resting comfortably, that dog should be evaluated for heart disease, respiratory disease, and other problems that can cause difficulty breathing. As MVD progresses, early symptoms which may occur are exercise intolerance, breathlessness, productive coughing, a distended abdomen, weight loss, and fainting. Breathlessness is a most common sign, starting as excessive panting on exercise. As breathing difficulties become more severe, the dog may sit or stand, holding its elbows away from the chest, and it may be reluctant to sit down.

As greater quantities of blood leak through the damaged mitral valve from the left ventricle back into the left atrium of the heart, the atrium gradually begins to swell to accommodate the overload of blood, and there is a reduction in the ability of the ventricle to provide sufficient

blood to meet the demands of the rest of the body. The heart then has to pump harder and faster, to meet those demands.

Also, due to the increasing lack of blood being pumped throughout the body, non-essential blood vessels begin to shut down, to conserve blood flow for vital organs, such as the brain and the heart itself, and reducing the flow to the skin and the kidneys. This causes the skin to pale and the kidneys to retain fluids in the circulation, because the circulation identifies the low cardiac output as dehydration. The shut-down of the distant blood vessels also has the effect of causing the left ventricle to beat against a higher resistance, causing another increase in mitral valve leakage. The excess fluid retention results in further stretching of the heart and greater mitral valve leakage, and the retained fluid is squeezed into other body tissues, the liver, chest, and belly.

The enlarged size of the heart can put pressure on the airway to the left lung, resulting in a dry, hacking cough and breathlessness. It may even cause the trachea to be compressed Fluid in the lungs can cause a soft moist cough, which may be followed by gagging up clear or pink tinged foamy fluid. Fluid accumulation around the heart and lungs simply causes difficulty breathing. When fluid accumulation is especially severe, the gums can turn gray or even blue from lack of oxygen.

Cavalier's heart failure 6 may display episodic weakness of the hindquarters, ataxia, or collapse, which is called presyncope, or combined with loss of consciousness, which is called syncope, due to a sudden decline in blood flow to the brain. Syncope is the medical term for fainting.

A loss of appetite, resulting in possibly severe weight loss (called cardiac cachexia), particularly of muscle mass, is another symptom of advanced MVD.

The ultimate consequence of mitral valve disease is heart failure. The median survival period for dogs once they develop severe congestive heart failure (CHF) due to MVD is approximately seven months, with 75% of the dogs dead by one year. For dogs with less severe CHF, the median survival period is one year, with 75% of the dogs dead by 21 months. However, the CKCS has a more accelerated version of MVD, and they typically progress more rapidly to heart failure.

Progression

The progression of mitral valve disease can be rapid or slow. Some Cavaliers develop a mild murmur without any more serious symptoms for many years. If the progression is slow enough, the dogs may die of other causes before their hearts reach failure. This is the usual pattern of MVD in most other breeds affected with it. In the Cavalier King Charles spaniel, some veterinary cardiologists have found prognostic value from the degree of mitral valve prolapse, the thickness of the leaflets, and whether ruptured tendinous chords are observed on the echocardiogram.

In most CKCSs, the disease shows a gradual progression in the loudness of the murmur and to more serious symptoms, in as little as 2 years after first detecting the murmur. Drugs may help to

minimize the symptoms, but eventually the drugs may be unable to control them. Severe symptoms in some Cavaliers will appear more quickly, although previously having been stable. If the tendinous chords rupture, and the valve leaflets cannot continue to open and close with each heart beat, death could be almost immediate.

Treatment

Preventative Vitamins and Supplements?

No medications or food supplements are known to prevent the onset of MVD. However, some supplements are intended to nourish the heart, although there is no scientific proof that they actually do so. The supplements below are known to be safe in dogs, and may be advantageous in Cavaliers.

- **Vitamin C** (300 to 400 mg. daily)
- **Vitamin E** (100 I.U. daily)
- **CoQ10** (30 mg. daily)
- Fish oils high in **Omega 3 Fatty Acids** (180 mg EPA per 10 pounds body weight)
- **Bio-Cardio** is a multi-vitamin, mineral, and herbal extract supplement made especially for dogs by Thorne Research.
- **Canine Cardiac** by Standard Process, is veterinary formulas to support heart function.
- **Cardio-Support** by D'Arcy Naturals, is veterinary herbal formula to aid heart function and blood circulation.

Holistic supplements should be taken only if prescribed by a licensed veterinarian who also is holistically trained.

Mild Murmur. A Cavalier with early mitral valve disease has a mild murmur but otherwise is symptom-free (asymptomatic). There may be minimal enlargement of the heart, as shown by x-ray or ultrasound scan. At this stage, there is no need for medical treatment, but heart size should be monitored by x-rays every 6 to 12 months. Overweight dogs should be put on a weight-reducing diet. Low salt diets have been suggested, to help reduce water retention. It would be prudent to avoid extreme exertion. If water in your area is high in sodium, bottled water would be advised.

A 2002 study sponsored by a drug manufacturer suggested that Cavaliers with only mild MVD murmurs and which otherwise are symptomless be prescribed angiotensin converting enzyme (ACE) inhibitors (**enalapril [Enacard, Vasotec], benazepril [Lotensin, Fortekor]**) as therapy to postpone or prevent congestive heart failure. However, a **2002 Scandinavian study** of asymptomatic Cavaliers with mild MVD murmurs has shown that the application of ACE inhibitors had no significant affect upon the time from the initiation of ACE inhibitor therapy to heart failure. At this time, we believe that dogs with MVD should be treated with ACE inhibitors only after it progresses to congestive heart failure.

Nutrition. General nutrition is very important. Cavaliers at this advanced stage may suffer severe weight loss, called progressive cardiac cachexia, and they should be fed palliative food to maintain muscle mass.

Moderate MVD. Moderate MVD is indicated by a louder murmur, increased breathlessness on occasions, occasional dry, hacking coughing, and moderate to severe enlargement of the heart on x-rays or scan, with some fluid present in the lungs. At this stage, reducing exercise will help to reduce the heart's workload. Treatment will be necessary at this stage, usually in a tablet form.

Diuretics. Since a dog with moderate MVD begins to retain fluid and salt, diuretics, which are drugs which cause the kidneys to excrete more fluid than normal, may be used to remove fluid from the lungs. Examples of diuretics include:

- **furosemide** – trade names **Lasix, Salix, Diuride, Frudix, Frusemide**
- **hydrochlorothiazide – trade name Dyazide**
- **spironolactone (Aldactone)**

The kidneys and electrolytes should be evaluated before starting diuretics and should be monitored periodically thereafter.

Natural diuretics include **Wu Ling San** and **Alisma**, both traditional Chinese herbal medicines (TCM). Other Chinese herbal alternatives include **Salvia Shou Wu**, a Seven Forests patented supplement which consists of Salvia extract, and several other herbs and flowers. **Parsley, corn silk and dandelion** are herbal diuretics. Holistic supplements should be taken only if prescribed by a licensed veterinarian who also is holistically trained.

ACE Inhibitors. ACE inhibitors block the angiotensin converting enzyme, which is necessary to produce a substance that causes blood vessels to tighten, are usually prescribed as soon as congestive heart failure develops. So, ACE inhibitors serve to relax the blood vessels (are vasodilators), thereby lowering the blood pressure and increasing the supply of blood and oxygen to the heart. ACE inhibitors include:

- **enalapril maleate - Enacard, Vasotec**
- **benazepril [Lotensin, Fortekor]**.

Recent studies have concluded that diuretics such as furosemide should be used only combined with ACE inhibitors, so that the diuretic dosage may be sharply reduced to avoid the worst of its negative side effects. As ACE inhibitors can lower blood pressure, blood pressure should be prior to prescribing these drugs, and should be monitored regularly. As well, kidney function should be monitored before and periodically after beginning ACE inhibitor therapy. ACE inhibitors do prolong survival in dogs with congestive heart failure, when used along with diuretics. However, in some cases, they are not well tolerated, and must be discontinued or used at a lower dose.

Hawthorn berry is an herb that has properties similar to ACE inhibitors. Dose of ACE inhibitors may need to be reduced if your pet takes a supplement containing Hawthorne. Holistic supplements should be taken only if prescribed by a licensed veterinarian who also is holistically trained.

Beta Blockers. Another drug being used by some veterinary cardiologists is **carvedilol (Coreg)**, a non-selective beta-blocker with anti-oxidant effects which reduces the heart's rate and the force of its contraction, thereby reducing the work of the heart. Carvedilol also causes the arteries to relax and the blood pressure to drop. Carvedilol is under study to determine its affect upon Cavaliers and other canines with asymptomatic MVD and whether it causes MVD to progress at a slower rate than dogs not taking the medication. Cavaliers with MVD who have weak heart contraction should probably not take Coreg. At this time, there is no strong evidence on which to recommend use of Coreg, and it is possible that it could make the heart contractions weaker, so we do not recommend it routinely for Cavaliers with MVD.

Severe MVD. Dogs with severe MVD will have difficulty breathing while at rest, and may not be able to tolerate even minimal exercise. Diuretics may be given by injection from time to time in severe cases. In addition to the medications for moderate MVD, dog with severe MVD may be treated with medications described below

Positive Inotropes are drugs which help a weak heart muscle beat with more strength. "fractional Shortening" as measure on an echocardiogram is a measurement of strength of heart muscle contraction. Positive inotropes include:

- **pimobendan - Vetmedin**
- **digitalis – Lanoxin, Cardoxin**

Texas A&M University Drs. Sonya G. Gordon, Matthew W. Miller, and Ashley B. Saunders find that "pimobendan is safe, well tolerated, and leads to enhanced quality of life in dogs with CHF secondary to...chronic valvular disease when used in combination with furosemide or other conventional therapies (e.g., angiotensin-converting enzyme inhibitors, digoxin)" and that"ongoing studies are evaluating its effects on mortality associated with chronic valvular disease

Digoxin a cardiac glycoside extracted from the foxglove plant (digitalis) may be used to improve heart muscle strength to attempt to help the heart contract more strongly, though its effect can sometimes be small. Pimobendan is a much more powerful drug to stimulate heart contraction.

Natural supplements which may help to strengthen and energize the heart of a dog with severe MVD include **D-Ribose**, also known as alpha-D-ribofuranoside, which reportedly improves ventilatory efficiency in patients with congestive heart failure (CHF).

Arterial Dilators. Drugs that dilate the arteries can help control fluid accumulation when diuretics and ACE inhibitors alone are unsuccessful. Arterial dilators include:

- **hydralazine**
- **pimobendan**
- **sodium nitroprusside**
- **sildenafil (Viagra)**

Viagra and similar drugs lower high blood pressure in the lungs (pulmonary hypertension), and are being prescribed when this is present. Doppler ultrasound is required in order to diagnose pulmonary hypertension. Pulmonary hypertension is often not present in dogs with MVD.

Bronchodilators. If there is no high blood pressure, some veterinarians may prescribe a bronchial dilator to help relax and open air passages in the lungs, making breathing easier. These drugs are used in only the most severe cases. Bronchodilators include:

- aminophylline
- oxtriphylline
- theophylline

Oxygen. In a crisis, oxygen therapy can make the difference between life and death. If can help get oxygen to the tissues while drugs have time to begin to work. Oxygen is most often given by placing the patient in an oxygen cage, which contains air with increase oxygen content.

Tapping the abdomen. If right heart failure develops, retained fluids in the belly (ascites), may need to be periodically removed by aspiration with a hypodermic needle. This procedure can improve breathing tremendously in some cases.

Mitral Valve Replacement Surgery. It is unrealistic to try to cure canine mitral valve disease. Replacement of the defective mitral valve is available at Texas A&M, but is still in the experimental stages at this point. Colorado State University's Veterinary Teaching Hospital has had a mitral valve replacement program for dogs since 1997. Owners should expect to pay at least $5,000-10,000 for this surgery. The program requires that the dog's kidneys and other vital organs be in ideal condition. Therefore, MVD typically is treated by managing heart failure. The goals of therapy are to improve the dog's quality and length of life.

Breeders' Responsibilities

Due to the pervasiveness of MVD in the breed worldwide, Cavalier King Charles spaniels under the age of five years should not be bred, in general. Also, no Cavalier

should be bred after age five years if it developed an MVD murmur before the age of five years. Any littermates of breeding stock having early-onset MVD (mitral valve murmurs before age 5 years) should be taken into very serious consideration for taking out of the breeding pool. All CKCS breeding stock should be examined by a veterinary cardiologist, or an internist who practices veterinary cardiology, at least annually. The closer the examination to the breeding, the better. It is recommended that all Cavaliers, breeding stock or not, be examined annually by a veterinary cardiologist after one year of age.

Support:

Two MVD support groups are Yahoo! Group: MVD in Cavaliers and Karlin Lillington's CavalierTalk: SM and MVD Cavaliers Forum.