

Congenital Heart Disease in Bull Terriers

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The congenital heart defects that have been identified in bull terriers are described below.

Congenital Heart Disease in Bull Terriers

Paper given to the
Combined Bull Terrier Clubs' Health Committee

by

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Overview of the normal anatomy of the heart

The heart has four chambers. The right atrium (RA) collects venous blood from the body, and it passes through the tricuspid valve to the right ventricle (RV). The right ventricle pumps the venous blood, through the pulmonic valve, into the pulmonary artery and so into the lungs, where the blood is oxygenated. The blood returns to the heart, into the left atrium (LA). Blood passes through the open mitral valve into the main pumping chamber of the heart, the left ventricle (LV). When this pumps, the mitral valve closes and blood is ejected out via the open aortic valve into the aorta, which divides to every artery to all the organs of the body.

Mitral Dysplasia

Mitral dysplasia is a malformation of the mitral valve apparatus. The normal mitral valve apparatus consists of two valve cusps. These are anchored by chordae tendinae to two papillary muscles in the left ventricle. The papillary muscles and the chordae allow the valve to close when the ventricle contracts, but restrain the valve so it does not flap back into left atrium. The closed valve should be a tight seal, allowing no blood into the left atrium.

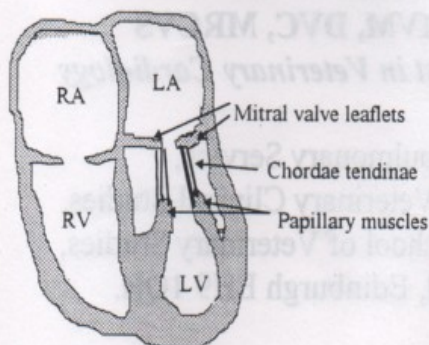


Figure 1

Diagram of the normal anatomy of the mitral valve apparatus

With mitral dysplasia in bull terriers, the mitral valve apparatus is deformed. The valve cusps become thickened and nodular. The papillary muscles are very large and thick,

and only very short, thickened chordae tendinae attach the papillary muscles to the valve leaflets.

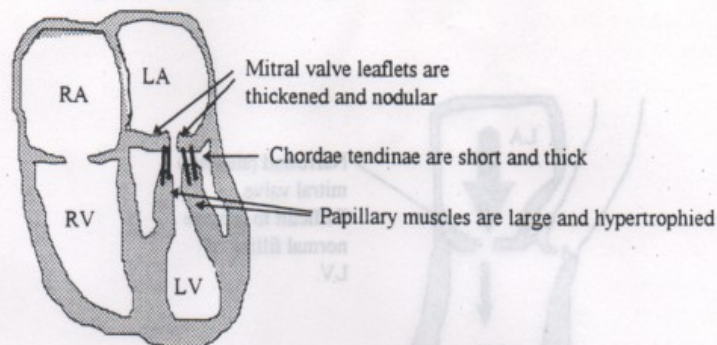


Figure 2:
Diagram of the abnormalities recognised affecting the mitral valve apparatus with mitral dysplasia.

The most common consequence of mitral dysplasia is a leaky (incompetent) mitral valve. During ventricular contraction (systole), instead of the mitral valve sealing shut, it allows a jet of blood to pass backwards into the left atrium, as well as pumping blood forward in the normal direction, into the aorta. This leak is known as *mitral regurgitation*. The turbulent jet of blood flow due to mitral regurgitation can be heard with a stethoscope - it is a *systolic heart murmur*, which is most intense over the mitral valve area of the dog's chest. Loud murmurs can radiate to other parts of the dog's chest as well.

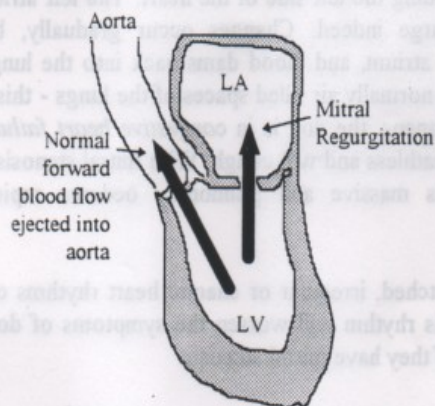


Figure 3.
Diagram of the left side of the heart and blood flow during ventricular contraction (systole):

- (i) normal forward flow into aorta
- (ii) backward flow across incompetent mitral valve (mitral regurgitation).

Occasionally, the mitral valve in mitral dysplasia can be narrowed as well, called *mitral stenosis*. It is difficult for the left atrium to empty into the left ventricle. Pressures can build up in the left atrium. The left atrium may become greatly enlarged.

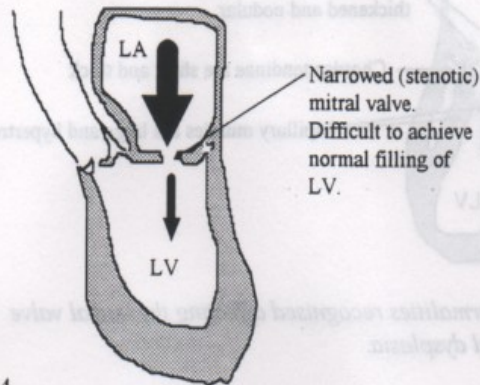


Figure 4.

Diagram of the left side of the heart showing ventricular filling (diastole) with mitral stenosis.

Consequences of mitral dysplasia

Bull terriers with mitral dysplasia will have a heart murmur. If the valve is only slightly leaky, the murmur is only soft and the dog may not be affected at all.

Where there is severe mitral regurgitation, a large volume of blood moves back and forwards across the mitral valve, overloading the left side of the heart. The left atrium and left ventricle can become very large indeed. Changes occur gradually, but eventually, pressures build up in the left atrium, and blood dams back into the lungs, and comes out of the circulation into the normally air filled spaces of the lungs - this is called *pulmonary oedema*. Once this happens, the dog is in *congestive heart failure*. Dogs with pulmonary oedema will be breathless and will cough. With mitral stenosis is also present, left atrial enlargement is massive and pulmonary oedema rapidly develops.

Where the left atrium becomes very stretched, irregular or chaotic heart rhythms can develop, such as *atrial fibrillation*. This rhythm will worsen the symptoms of dogs with congestive heart failure, especially if they have mitral stenosis.

Dogs with severe mitral dysplasia have poor heart output into their circulation - they may faint on exertion or excitement or stress (*syncope*) as their brains can become short of oxygen.

Aortic Stenosis

The aortic valve is positioned between the left ventricle and the aorta. It opens as the left ventricle contracts, allowing the left ventricle to eject blood into the aorta. It normally offers no restriction to blood flow.

With aortic stenosis, the aortic valve itself or a fibrous band beneath the valve (subaortic stenosis) restricts the outflow to blood from the left ventricle. The left ventricle has to work hard just to eject blood, and the heart muscle becomes thickened (hypertrophied) to compensate. Blood ejecting past the obstruction to outflow is very fast and turbulent. This turbulent flow causes a heart murmur which can be detected with a stethoscope over the aortic valve area of the chest wall. The louder the murmur, the more severe the aortic stenosis and the faster and more turbulent the blood flow passing the obstruction. This turbulent blood flow can affect the walls of the aorta, and it can stretch, called post-stenotic dilatation.

As the left ventricular muscle thickens, it exceeds the coronary artery blood supply to it. This means that areas of the heart muscle can become deprived of oxygen. The muscle becomes irritable and may cause abnormal beats (called *ventricular premature complexes or VPCs*). A run of these abnormal beats (*ventricular tachycardia*) can cause the dog to faint or even to suddenly die.

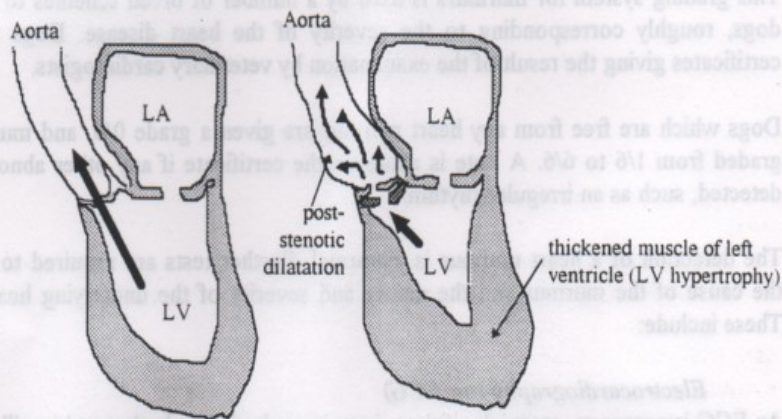


Figure 5a.
Diagram of the left side of the heart showing normal unrestricted outflow from the left ventricle to the aorta through the aortic valve.

Figure 5b.
Diagram of the left side of the heart with severe aortic stenosis. A fibrous band restricts outflow from the left ventricle into the aorta. The left ventricular muscle becomes thick as it works to produce sufficient pressure to pump the blood into the aorta. Blood flow is fast and turbulent as it enters the aorta, and the aorta can stretch (post-stenotic dilatation).

Aortic stenosis or subaortic stenosis does occur in bull terriers. However, it is more common in association with mitral dysplasia. Dogs with both defects are very severely affected, particularly if they have mitral stenosis as well.

Diagnosis of congenital heart disease in bull terriers.

Examination by a veterinary cardiologist

As explained above, congenital heart disease is manifested by a heart murmur, which is detected by a veterinary surgeon or a veterinary cardiologist by careful clinical examination, including auscultation, with a stethoscope. Because bull terriers are not easy dogs to auscultate, sometimes the dog's own veterinary surgeon may not have detected the murmur.

Heart murmurs show that there is turbulent blood flow in the heart, which is abnormal. Murmurs are graded out of 6 - where grade 6 is the loudest murmur which can be detected even without a stethoscope, with a "thrill" on the dog's chest wall overlying the heart. Grade 1 murmurs are the most quiet and difficult to detect. A very quiet room, a good quality stethoscope and a quiet, calm dog who is not panting are required before this murmur is detected.

What is "heart murmur scoring"?

This grading system for murmurs is used by a number of breed schemes to score the dogs, roughly corresponding to the severity of the heart disease. Dogs are given certificates giving the result of the examination by veterinary cardiologists.

Dogs which are free from any heart murmur are given a grade 0/6, and murmurs are graded from 1/6 to 6/6. A note is made on the certificate if any other abnormality is detected, such as an irregular rhythm.

The detection of a heart murmur is abnormal. Further tests are required to diagnose the cause of the murmur and the nature and severity of the underlying heart defect. These include:

Electrocardiography (an ECG)

An ECG is necessary especially if there is an irregular heart rhythm and it will show the veterinary cardiologist what treatment may be necessary to control the rhythm. The ECG may also indicate whether certain heart chambers are enlarged. The ECG records the electrical activity from the heart with painless electrodes attached to the loose skin of each leg.

Radiography (X rays)

Where heart failure is diagnosed or suspected, chest X rays will show how much fluid has accumulated on the lungs. The enlargement of the heart chambers is also shown. X rays are a specific way to show how overloaded the heart may be. Usually, the dog is sedated before X rays are taken, so he lies on the table preventing unnecessary X ray exposure of the vets, staff and owner. Obviously, the sedation and dose rates are selected to be safe even for dogs in heart failure.

Echocardiography and Doppler

Echocardiography is an ultrasound scan of the heart. It is similar to the scan in pregnancy, checking the baby is healthy. It is completely safe and does not even require a sedative or anaesthetic. The heart is seen in real time actually beating and each chamber and each heart valve can be seen. Measurements of contractility and other parameters of heart function can be made. Doppler echocardiography is a special

form of ultrasound, where the speed and character of blood moving through the heart can be recorded - as a spectrum or even in colour, where colour flow echocardiography is available. Abnormal jets of blood due to leaky or narrowed valves can be measured, which allows the cardiologist to determine how severe the heart disease is, so to provide appropriate advice to the owner and treatment to the dog. Doppler echocardiography is only available at certain cardiology referral centres as the equipment is very expensive and considerable expertise is required to perform and analyse the scan.

Blood samples

These may be taken from a dog with heart disease, especially when they are in heart failure. It is important to check on kidney and liver function before prescribing certain drugs. During treatment, the vet may want to check the blood level of certain drugs.

It is very important to investigate heart disease, so that the best decisions for advice and treatment of each individual dog can be made.

Treatment of bull terriers with heart disease

Obviously, the treatment will vary depending on the diagnosis and the severity of the underlying heart disease.

Dogs with heart failure with mitral dysplasia will require diuretics, to remove excess fluid from the lungs. Drugs called ACE inhibitors (angiotensin-converting-enzyme inhibitors) such as Cardiovet^R (enalapril) or Fortekor^R (benazapril) are quite expensive, but they counteract some of the adverse hormones which are elevated in heart failure. They have been shown to prolong life and improve quality of life of humans and dogs with heart failure. However, they may be dangerous in bull terriers with mitral stenosis or aortic stenosis - which shows the importance of a complete investigation of the actual diagnosis of the heart disease. Other drugs may increase the contractility of the heart and control any abnormal heart rhythms.

A specialist cardiac surgeon from the Royal Veterinary College in London has recently replaced an abnormal mitral valve in a bull terrier. If this is considered, it must be done before any heart enlargement occurs or the dog develops heart failure.

How do we control heart disease in bull terriers?

Modern veterinary cardiology and advances in cardiac surgery can help individual dogs with heart disease and heart failure. Much more important for the future of the bull terrier breed, however, is to prevent heart disease. Certain questions need to be answered:

(i) How common is heart disease in bull terriers?

The author has examined a large number (over 200) of bull terriers since 1989. Since then, about 30% of all bull terriers screened have a heart murmur. These are mainly show dogs - likely to be the breeding stock of the future.

About 80 bull terriers with heart murmurs have received full cardiac investigation by the author and colleagues. Most of these dogs have mitral dysplasia, with a few having mitral stenosis or aortic stenosis.

With the formation of the Combined Bull Terrier Clubs' Health Committee, individual breed clubs should be encouraged to have a veterinary cardiologist to screen bull terriers at their open or championship shows. A veterinary cardiologist is a veterinary surgeon who has the RCVS Certificate in Small Animal Cardiology (CertSAC), or Certificate in Veterinary Cardiology (CertVC), or the Diploma in Veterinary Cardiology (DVC). The use of these veterinary surgeons rather than the dogs' own vet means that a known standard of screening is achieved. A certificate is completed by the cardiologist (example copy enclosed). Results should be collated by a single person - possibly a retired breeder, or at least someone that all club's members will trust. All information received should be absolutely confidential. By this method, the Health Committee is able to ascertain the prevalence and significance of heart disease in all geographical areas. By recording the sire and dam of each dog and from screening extended families, pedigree analysis may be used to try and determine the mode of inheritance of heart disease in bull terriers.

Dogs with significant heart murmurs (grade 2/6 or louder) should receive further cardiac investigation to determine the nature of the underlying heart disease, as described above.

(ii) *Is heart disease inherited in bull terriers?*

This is not yet proven. However, the fact it is common in the breed, and in certain family lines, there is a significant clustering of heart disease, it probably is inherited. Careful family history, cardiac examinations and pedigree analysis of affected bull terrier families is necessary to prove this.

(iii) *My dog had a heart murmur and lived until he was 13!*

Many bull terriers owners and breeders have said this to me! Some bull terriers may have a heart murmur due to mild mitral dysplasia or aortic stenosis and lead a normal life and have a normal life span. This is great for that dog, but his offspring may not be so lucky. The defect may be more severe and many puppies may die suddenly, or develop heart failure. It is important that bull terrier breeders recognise the genetic importance of a dog or bitch with a heart murmur to consider the well being of the breed as a whole.

(iv) *Setting up a heart testing scheme*

Once the Combined Bull Terrier Club's Health Committee has screened a large number of dogs and the prevalence of heart disease is known, some recommendations can be made. From the author's initial results, where about 60+% of dogs tested are free from heart murmurs, it may be possible to advise only breeding from dogs which have no evidence of heart disease and are heart murmur free (0/6). Obviously, in selecting a dog as a sire, or bitch as a dam, there are many things which need to be considered, including temperament and various health matters and a balance is necessary. Certainly, veterinary cardiologists would advise that no dog or bitch is used for breeding with a grade 2/6 murmur or louder. Members of the committee and the various bull terrier clubs may decide to allow names of dogs and bitches to be published where they have no heart murmur, which will help breeders wanting to select away from heart disease. The progeny of matings should also be tested and the results

analysed as part of the pedigree analysis. One hopes that where a sire and dam with no heart murmurs are bred, the offspring should also be free from heart disease!

Conclusion

It is a devastating thing to watch a young bull terrier, who should be in the prime of life, slowly die with heart failure or even drop dead suddenly. Breed club schemes with other breeds, such as the Boxers and Newfoundlands in the UK have helped greatly reduce the incidence of heart disease and the severity of heart disease in these breeds, although there are on-going teething problems and it is not easy for breed clubs to take this on. It is the only way we can protect bull terriers in the future from being born with heart disease. Veterinary cardiologists in the UK are very impressed with the foresight of the Combined Bull Terrier Clubs Health Committee and we encourage you to consider setting up this scheme carefully.

Discussion

This paper attracted a number of carefully considered questions, which Mrs Dukes McEwan answered very efficiently. Asked 'Has our breeding changed the physiology of the dogs and therefore the incidence of heart disease?' she replied that this was possible, but there was really no way of knowing. Much interest focused on the desirability of testing dogs at dog shows, where a specialist in veterinary cardiology could most efficiently deal with a substantial number of dogs. The speaker considered that a show venue should be suitable despite the excitement associated with shows, attending a veterinary surgery as an alternative was probably equally exciting for most dogs. Asked about the subjective nature of the testing she agreed that it was indeed subjective, which was why a specialist in this field was essential. She was able to suggest to show managements who wanted them, specialists in various parts of the country. Dealing with a question on the grade of heart murmur at which dogs might be expected to show symptoms, she suggested grade 3, but it also depends on the type of heart disease. One questioner wanted to know the ideal age for heart testing, and was told that a dog of normal show age, say six months to two years was suitable. Old dogs might of course develop heart disease due to non-heritable causes. A question on whether a heart murmur could be affected by a bitch being in season was answered in the affirmative - possibly because of changes in blood viscosity associated with oestrus.

Perhaps the most fundamental question related to the mode of inheritance. Mrs Dukes McEwan said that she suspected that the Bull Terriers' mitral dysplasia was inherited, but pedigree analysis was required to determine the mode of inheritance (some other congenital heart disease in other breeds is suspected to be autosomal dominant with variable penetrance - such as aortic stenosis in Newfoundlands). She had collected a considerable quantity of information and associated pedigrees which might throw some light on this subject, but had been unable so far to devote sufficient time to a full analysis. Finally, the answer to "Would you be interested in the eventual autopsy of tested dogs which had been found to have a heart murmur?" was "Yes, it would advance our knowledge."

