Mitral Valve Dysplasia in Eight English Springer Spaniels

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Running title: Mitral Dysplasia in English Springer Spaniels

**Abstract**

**Introduction:** To describe the signalment, physical examination and echocardiographic findings of a series of English Springer Spaniels (ESS) diagnosed with congenital mitral valve dysplasia (MD).

**Animals:** Eight client-owned ESS with congenital MD referred for murmur investigation and/or suspected congestive heart failure (CHF).

**Materials and Methods:** Retrospective case series. Medical records and echocardiograms were reviewed to collect relevant data. Echocardiograms were assessed for the following abnormalities consistent with MD: thickened valve leaflets or leaflet tips, a ‘hockey stick’ appearance to the valve leaflets, abnormal length of one leaflet with respect to the other, tethering of one or both leaflets to the ~~interventricular septum or~~ papillary muscles

**Results:** All eight dogs showed the typical echocardiographic lesions associated with MD: thickened leaflet tips (5/8), ‘hockey stick’ appearance (5/8), elongated anterior leaflet (4/8), tethering of one or both leaflets (7/8). Seven of the eight dogs presented in CHF. Six of the 8 dogs had left ventricular dilation in both systole and diastole. Two of the eight dogs had reduced systolic function as assessed by ejection fraction/fractional shortening, however end-systolic volume index was increased in 6/8 dogs. Two dogs subsequently developed atrial fibrillation.

**Conclusions:** Congenital MD should be considered in ESS with a left-sided apical systolic murmur, particularly in younger dogs. The valve changes seen are similar to those reported in other breeds with MD (thickened leaflet tips, hockey stick appearance to open leaflet tips, abnormal leaflet tethering, abnormally shaped leaflets) and may result in marked remodeling and CHF.

**Keywords:** Canine; congenital heart disease; echocardiography

Abbreviations

|  |  |
| --- | --- |
| AF | atrial fibrillation |
| CHF | congestive heart failure |
| DCM | dilated cardiomyopathy |
| EF | ejection fraction |
| E/IVRT  | ratio of mitral E wave velocity to LV isovolumic relaxation time |
| ESS | English springer spaniel |
| EDVI  | end-diastolic volume index |
| ESVI  | end-systolic volume index |
| FS | fractional shortening |
| LA  | left atrium |
| LV | left ventricle |
| MD | mitral dysplasia |
| MMVD  | myxomatous mitral valve disease |
| MR  | mitral regurgitation |
| MS | mitral stenosis |
| MV | mitral valve |
| PDA | patent ductus arteriosus |

Introduction

Mitral valve dysplasia (MD) is an uncommon canine congenital condition, representing approximately 2-8% of congenital cardiac defects [1,2]. It is characterized by one or more of the following lesions seen on echocardiography: thickened mitral valve (MV) leaflets, often most notably at the leaflet tips, a hockey stick appearance to the leaflet tips, which are turned inwards during diastole (diastolic doming), abnormally long or short leaflets, abnormal chordae tendineae (usually shortened, thickened; sometimes elongated), tethering of the valve leaflets to the papillary muscles and malformed papillary muscles [1,3,4]. As a consequence the valve motion, as assessed by echocardiography, is often abnormal [4]. The presence of mitral regurgitation (MR) is variable and may result in volume-loading of the left heart, i.e. eccentric left ventricular (LV) hypertrophy and left atrial (LA) dilation, which may eventually result in increased left sided filling pressures [4]. Great Danes, English bull terriers, German shepherd dogs and Dalmatians are predisposed to MD [5-9].

Concurrent mitral stenosis (MS) is uncommonly seen in dogs

 and appears to be associated with a poorer prognosis [10]. It appears to be a significant concern in English bull terriers [6]; no other breeds appear to be predisposed, although in one study Newfoundlands made up 5/15 dogs with MS [10].

The disease progression and outcome of MD is variable, with some dogs developing clinical signs and congestive heart failure (CHF) from a young age, while others may remain asymptomatic [1,5-10].
English springer spaniels (ESS) are not commonly reported to be affected by any specific cardiac disease although in one study, five were diagnosed with atrial myopathy/persistent atrial standstill [11]. A genetic mutation associated with QT prolongation has also been identified in a family of ESS [12]. They may be predisposed to dilated cardiomyopathy (DCM) [13], though recent publications have refuted this [14]. They do not appear to be predisposed to myxomatous mitral valve disease (MMVD) based on the veterinary literature although this may reflect the inherent selection bias in most MMVD studies as it is typically a disease affecting small breed dogs [15,16], so ESS may have been excluded.

The purpose of this report is to describe the clinical features of MD in ~~14~~ eight ESS that presented to a referral hospital.

Animals, Materials and Methods

This retrospective study obtained ethical approval from the local Veterinary Research Ethics Committee (reference: VREC: 632).

Electronic medical records from the record management systema of the Small Animal Teaching Hospital, University of Liverpool from 2010-2018 were reviewed to identify ESS with an echocardiographic diagnosis of MD. Additionally, echocardiogramsb from ESS diagnosed with MMVD and/or DCM were re-evaluated for features of MD as both diseases may present with mitral regurgitation and, in the case of MMVD, valvular pathology. Only ESS aged <5 years were included in this study, to reduce the likelihood of inadvertently including a dog with acquired MMVD or DCM.

For each dog, recorded patient data included signalment, historical information, presenting complaint, results of physical examination, results of diagnostic tests (~~troponin I,~~ electrocardiogram, echocardiography, thoracic radiographs), medications at the time of first presentation and those treatments prescribed on discharge.

All dogs had transthoracic echocardiography performed by a board-certified cardiologist or cardiology resident under the direct supervision of a board-certified cardiologist. Recorded echocardiographic measurements included LV internal dimension, actual and indexed for body weight, in diastole and systole from M-mode measurements of the LV in short axis, using leading edge to leading edge method [17]. Fractional shortening (FS) was calculated. Simpson’s method of discs was used to determine LV volume on images acquired from the right parasternal 4-chamber view optimized for LV length and area, including the LV apex. End-diastolic and end-systolic volume, indexed for body surface area (EDVI and ESVI, respectively); obtained by the formula: body surface area = (10.1 x [body weight in kg]0.67)/100), and ejection fraction (EF) were calculated. Mitral E-point to septal separation was measured from M-mode images taken from the right parasternal short axis view at the level of the MV. Transmitral Doppler E-wave velocity and A-wave velocity were measured using pulsed-wave Doppler from the left apical four-chamber view, aligned for mitral inflow with the sample volume between the MV leaflet tips; from these the mitral E/A ratio was calculated. From a left apical five-chamber view showing color of mitral inflow and LV outflow, with the sample volume placed in the red/blue interface, the isovolumic relaxation time (IVRT) was measured and the ratio of the mitral E/IVRT calculated as an estimate of left-sided filling pressures [18]. Echocardiographic measurements were compared with reference ranges published for ESS [14]. The echocardiograms were retrospectively reviewed by two authors (SS and JHE) for specific echocardiographic features of MD and MS. For inclusion in the study a dog had to show echocardiographic evidence of any two of the following signs of dysplastic MV apparatus (with agreement between both observers), with resultant MR: thickened leaflets and/or leaflet tips, a ‘hockey stick’ appearance to one or both of the open MV leaflets during diastole (also called diastolic doming); abnormal length of one leaflet with respect to the other; or tethering of one of the leaflets to the papillary muscles. The valves were considered to be tethered if one or both leaflets were clearly attached to the papillary muscles in such a fashion that it interfered with normal valve movement (closure or opening).

Valve motion was also assessed for the presence of prolapse or tenting during ventricular systole. Tenting of the MV was defined as failure of the MV leaflets to coapt at the level of the mitral annulus in systole, resulting in a ‘tented’ appearance to the area between the mitral annulus line and the closed MV leaflets. This feature would not be expected to be seen with MMVD, in which systolic prolapse is commonly reported [16, 19]. Valve motion was assessed on either the right parasternal long axis four chamber view or the left apical four chamber view. Tenting was not used as a diagnostic criterion for the presence of MD as it is not specific, but rather occurs as a result of LV remodeling and altered geometry (increased sphericity) of the LV [20].

Echocardiograms were assessed for the presence of concurrent MS based on the presence of one or more of the following criteria, as previously described [6, 10]: thickened MV leaflets with concordant anterior and posterior leaflet motion on mitral M-mode; reduced E-F slope on mitral M-mode; or prolonged E wave deceleration with increased PHT on pulsed or continuous wave Doppler interrogation of the transmitral inflow. Pressure half time was measured as the time taken for the transmitral pressure gradient to decrease by half, as previously described [10]. Normal PHT was defined as <50ms [21], although a breed-specific value for ESS is not available.

A diagnosis of left-sided CHF was made based on clinical signs, the presence of pulmonary venous congestion and lung infiltrate consistent with cardiogenic pulmonary edema on thoracic radiographs (reviewed by a board-certified diagnostic imager or diagnostic imaging resident under the direct supervision of a board-certified diagnostic imager) and/or a positive response to diuretic therapy if this had been instigated prior to investigations.

All qualitative and quantitative data was collated into spreadsheetsc. Basic descriptive statistical analysis included visual inspection of data and the Shapiro-Wilk test to identify whether data was normally distributedd. Vertebral heart score was normally distributed and is presented as mean (± standard deviation). No other data were normally distributed; as such the variables are presented as median with range (minimum – maximum).

Results

Signalment and Clinical Presentation

Fifty-five ESS had echocardiograms performed between 2010 and 2018. Of these dogs, 12 were initially diagnosed with MD, six of which were <5 years old at the time of diagnosis and thus eligible for inclusion in the study. Two other dogs were initially diagnosed with MMVD and DCM (one each) but were reclassified as MD following re-evaluation of the echocardiogram, resulting in a total of eight dogs meeting inclusion criteria.

Signalment and ~~descriptive statistics~~ clinical information for the eight ESS are presented in the supplementary table (available online). The study group comprised six males and two females. The median age was 27.5 months (range: 4 – 54). The median body weight was 15.8kg (range: 11.4 – 20.4). Two patients presented asymptomatically for murmur investigations whilst the remaining patients presented with a combination of clinical signs (NB: Some dogs displayed more than one clinical sign): tachypnea/dyspnea (3 dogs), lethargy (5 dogs) and/or cough (4 dogs). Three dogs had received cardiac treatment at the time of presentation including furosemide (3 dogs); benazepril (2 dogs); pimobendan (1 dog) and spironolactone (1 dog). In all cases the furosemide had been administered in the immediate period prior to referral (five days at most).

Seven dogs had a left apical systolic heart murmur identified ranging in intensity from III-V/VI. One dog had a left basilar continuous murmur of intensity V/VI (the patient had a concurrent patent ductus arteriosus (PDA)). The findings are summarized for each dog in the supplementary table (available online).

Echocardiographic Findings

All dogs had complete echocardiographic reports and images available for review. All dogs had multiple morphological abnormalities of the mitral valves; 5/8 had thickened valve leaflet tips (figure 1A, videos 1 and 2), 4/8 had a ‘hockey stick’ appearance to one or both leaflets during diastole (figure 1B, videos 2 and 5), 4/8 had an abnormally elongated anterior valve leaflet, compared to the posterior leaflet (figure 1A, 1C, video 3). Tethering of one or both leaflets to the ~~interventricular septum or~~ papillary muscles was seen in 7/8 dogs; 5/8 had tethering of the posterior leaflet (Figure 1A, 1C, videos 4 and 5), 1/8 had tethering of the anterior leaflet and 1/8 had tethering of both leaflets. All dogs had mitral regurgitation as a consequence of MD (video 4). Seven dogs had a tented appearance to the valve apparatus during ventricular systole (figure 1D, figure 2, video 6). No dog in this series had evidence of concurrent mitral stenosis, based on assessment for concordant valve leaflet motion, reduced E-F slope on M-mode or pressure half time on pulsed/continuous wave Doppler interrogation of transmitral inflow.

The median LV diameter in diastole was 60.5mm (range: 38.0 - 69.8mm) and in systole was 50.0mm (range: 27.6 – 61.2mm). Left ventricular dilation based on M-mode measurements using published ESS reference ranges was evident in 6/8 dogs both in systole (>38.8mm [14]) and diastole (>48.5mm [14]). The LV dimensions were also normalized for body weight and compared with published intervals [17]; by these criteria 7/8 dogs had LV dilation both in systole (>1.26 [17]) and diastole (>1.85 [17]). Median sphericity index was 1.1 (range: 0.9 – 1.6); sphericity index was reduced (<1.23 [14]) in 5/8 dogs. Median EDVI was 216.7mL/m2 (range: 56.7 – 296.9mL/m2); increased EDVI (>114 mL/m2 [14]) was found in 6/8 dogs. Median ESVI was 104.3mL/m2 (range: 47.3 – 201.4mL/m2); increased ESVI (>68.1mL/m2 [14]) was found in 6/8 dogs. Median FS was 22.5% (range: 7.0 – 28.0%); reduced FS (<17.3% [14]) was found in 2/8 dogs. Median EF was 45.1% (range: 16.3 – 60.6%); reduced EF (<36% [14]) was found in 2/8 dogs. Median mitral E-point to septal separation was 10.3mm (range: 1.6 – 32.5mm); three dogs had increased mitral E-point to septal separation (>10.5mm [14]).

Thoracic Radiographic Findings

All dogs had thoracic radiographs obtained on presentation. All had evidence of cardiomegaly; mean vertebral heart score was 13.2 ± 1.2. Pulmonary venous congestion was reported in 4/8 dogs (including the dog with a PDA who also had evidence of pulmonary arterial distention (reflecting pulmonary overcirculation)). An interstitial-to-alveolar lung pattern consistent with pulmonary edema was reported in 6/8 dogs. One dog did not have clear evidence of pulmonary edema but had received five days of diuretic therapy prior to investigations and had marked cardiomegaly and increased left-sided filling pressures seen on echocardiography; as such 7/8 dogs were diagnosed with CHF.

Arrhythmias

Two dogs developed atrial fibrillation (AF) four and five months after initial presentation; this was diagnosed during a routine follow up examination by six lead ECG. One other dog had occasional isolated ventricular premature complexes noted on the simultaneous ECG performed during echocardiography; further investigations were not performed.

Treatment

All seven dogs presenting in left-sided CHF received standard therapy including furosemide, pimobendan, benazepril and spironolactone. One dog subsequently received torasemide and hydrochlorothiazide due to refractory CHF. Both dogs subsequently diagnosed with AF received a combination of digoxin and diltiazem. The dog that did not present with signs of CHF received no treatment initially but was prescribed pimobendan on follow-up visits due to further cardiac remodeling.

Outcome

At the time of writing, 3/8 dogs had been euthanized due to worsening CHF with all of these dogs originally presenting in left sided CHF and including the two dogs that developed AF. The time to death ranged from 119-749 days from initial presentation. The remaining five dogs were alive, with a follow-up time ranging from 394-2007 days.

Discussion

To the authors’ knowledge, this is the first time MD has been described in ESS in the veterinary literature. The MV changes seen in this population are similar to those seen in other breeds with MD described [1,4,9]. No single abnormality was seen in all cases, however thickened valve leaflet tips with a ‘hockey stick’ appearance predominated in this study population.

English Springer Spaniels have not been previously reported to be predisposed to MD, or indeed any other congenital cardiac disease. There were no ESSs in one large retrospective analysis of congenital defects [2], and the population studied in another large paper by Tidholm et al. (1997) had only one ESS, with endocardial fibrosis [1].

It may be that ESS are less popular in mainland Europe and the USA compared with the United Kingdom, which may in part explain their absence from other studies. Additionally these populations may have genetic differences that could contribute to disease development; further studies are indicated in this regard.

Morphologically, MD can be misdiagnosed as MMVD which presents with similar abnormalities including MR, in the presence of increased thickness of one or both MV leaflets with the lesions often progressing over time [19]. It should be noted that large-breed dogs with MMVD tend not to have such marked valvular nodular thickening as small-breed dogs [19]; the valve is usually thin, elongated and prolapsed, although other authors note that the prolapse is less severe than in small-breed dogs [19]. No dogs in our study population showed valve prolapse, rather tenting was more common, seen in in 7/8 dogs. Prolapse is not described in the literature describing MD [5,9]. This is in keeping with findings from human literature; people with valve prolapse do not show restrictive opening of the MV leaflets during diastole, whereas restricted valve motion was a common finding in our patients, due to chordal/papillary muscle abnormalities [22].

English Springer Spaniels do not feature significantly in the published literature regarding large-breed dogs with MMVD; indeed, from five large MMVD studies in which the inclusion criteria allowed large-breed dogs, only five ESS featured from a total of 602 dogs [23-27]. More recently, only 2/42 ESS in a study by Dickson et al. were reported to have MMVD [14]. However, 24/42 dogs had trace/mild MR with normal valve morphology; the significance of MR in these dogs is unclear in the absence of morphological valve changes.

One dog in this study was initially diagnosed with MMVD however on subsequent revisits was reclassified as MD (video 3). This patient had thickening of the MV leaflet tips, particularly the anterior leaflet, however upon re-examination it was felt that the anterior leaflet elongation and suspected diastolic doming was more in keeping with MD, as was the absence of valve prolapse.

As MMVD is an acquired, degenerative disease commonly documented in older canine patients, MD should be primarily suspected in a younger dog presenting with MV abnormalities [7]. This patient was an adult at the time of diagnosis (54 months) and the valvular changes were not as dramatic as those seen in more severely-affected individuals; this highlights the possible difficulty in distinguishing between the two conditions. One could argue that in terms of prognosis and therapy the differentiation between MD/MMVD is immaterial in adult dogs (unless concurrent MS is present); however identifying congenitally affected individuals may have implications for breeding.

One dog in this study was originally misdiagnosed as having DCM (Videos 4 and 5), but re-examination of its echocardiographic images, combined with the young age (8 months) led to morphological reclassification as MD. English Springer Spaniels have historically been reported to be predisposed to DCM [13], however do not appear in large numbers in the DCM studies in the veterinary literature; 3/62 dogs in one study [28] and 4/369 in another [29]. Recently, Dickson et al. (2016, 2017) reported that healthy ESS often have rounded hearts and low systolic function indices, which may lead to some dogs being misclassified as having DCM [14,30]. Particularly fit ESS have also been reported to have lower systolic function parameters than less fit dogse; the athletic capability of the dogs in this study is not known, however may also be a factor. It may be difficult to distinguish between MD and DCM, particularly in older patients with marked cardiac remodeling and systolic dysfunction. It has been found that large-breed dogs with MMVD have reduced systolic function [19]; it is not unreasonable to consider that this may therefore be a feature of MD in large-breed dogs as well, although this has not been reported in the veterinary literature. Based on the diagnostic criteria set out by Dukes-McEwan et al. (2003) one would expect FS at least <20% and/or EF at least <40% in addition to increased sphericity [31]. In ESS the cut-off may be even lower given the reference intervals for normal FS and EF are 17% and 36% respectively [14]; only 2/8 dogs had values below this reference range. It should be noted, however, that in the presence of significant MR, FS and EF will be increased due to the reduced LV afterload; therefore, these measurements may be less reliable if there is moderate to severe MR. End-systolic volume index has been suggested as a more accurate measure of systolic dysfunction in dogs with significant MR [32]; given the similar LV loading conditions of MD, the finding of an increased ESVI in most (6/8) dogs is consistent with impaired systolic function. For these reasons it must be stressed that the diagnosis of MD should be made on the presence of multiple abnormalities: changes such as valve thickening, ‘hockey stick’ appearance, leaflet elongation and/or tethering suggest a primary valvular abnormality and would be more supportive of MD than DCM.

Ultimately, DCM is a diagnosis of exclusion; the presence of valvular pathology that could lead to the development of LV dilation precludes the diagnosis of primary (or idiopathic) DCM [31]. In DCM, the valves are morphologically normal but patients often have functional MR due to altered geometry of the LV.

Tenting of the MV leaflets may be seen in both DCM and MD. In human medicine it is reported that tenting can occur secondary to LV remodeling both in ischemic and non-ischemic MR [20,33,34], the latter being analogous to canine primary DCM. This results in rounded LV geometry, abnormal alignment of papillary muscles and chordae tendineae with dilation of the MV annulus, poor coaptation of the valves and a ‘tented’ appearance [20,33]. Patients with MD may have primary abnormalities of valve apparatus as well, such as leaflet tethering which may contribute to tenting. Tenting of the valve leaflets occurs to a degree in normal dogs, with variability documented between breeds [35] and differentiating between normal and pathological tenting is somewhat subjective. As such, it cannot be used solely as a diagnostic criterion for MD and further studies are indicated to evaluate severity of tenting in different disease processes.

One dog in this study also had a PDA, which may have contributed to cardiac remodeling and MR (and the pulmonary overcirculation seen on radiographs). This may also have contributed to valve tenting, as described previously. However, morphological changes to the MV were present, suggesting concurrent primary pathology. This patient had an Amplatz® Canine Duct Occluderf implanted; following the procedure there was a reduction in the LA and LV dimensions despite persistence of severe MR due to the MD.

Ultimately, a diagnosis of MD may be challenging in the presence of concurrent cardiac disease, however it should be suspected when characteristic valvular lesions are present, particularly in young dogs.

Two dogs subsequently developed AF. Atrial fibrillation has been found to be more common in large-breed dogs with degenerative MMVD than small-breed dogs [19], most likely due to the larger left atrial size [36, 37]. Atrial fibrillation is known to be a negative prognostic indicator in dogs with MMVD [27]; due to low numbers survival analysis was not calculated in this group, although it seems reasonable to assume AF would be similarly detrimental to dogs with congenital MD.

There are veterinary case reports describing successful outcomes for dogs with MD treated by surgical valve repair or replacement [38,39,40]. Given the increasing safety and availability of surgical valve repair [41,42], early detection and correct diagnosis of MD in ESS may result in better outcomes for affected dogs.

This study has limitations that must be addressed. As the study was retrospective, there was no set diagnostic protocol in these patients and treatments were administered at the discretion of the responsible clinician (or the referring veterinary surgeon). This was a small study population in dogs that presented to a referral hospital in the North West of England; the findings therefore may not represent larger ESS populations in the United Kingdom or other parts of the world. The diagnosis of MD was based primarily on subjective assessment of valvular changes and the study was not blinded, so biases may be present. Furthermore, the published ESS echocardiographic reference ranges are from dogs aged 2.5 years or older; four of the dogs in this study were younger than this and the ‘normal’ reference ranges may be less accurate. However, it should be considered even more abnormal for these younger individuals to fall outside the breed references for adult dogs.

Conclusions

Congenital MD should be considered in ESS with a left-sided apical systolic murmur, particularly in younger dogs. The valve changes seen are similar to those reported in other breeds with MD (thickened leaflet tips, hockey stick appearance to open leaflet tips, abnormal leaflet tethering, abnormally shaped leaflets) and may result in marked remodeling and CHF. In this small population of dogs, the rate of disease progression and outcome was highly variable; further studies are indicated to better evaluate these factors.

Conflict of Interest:

The authors do not have any conflicts of interest to disclose.

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| Case | Gender | Weight (kg) | Age at dx (m) | Murmur (/6) | Arrhythmias | CHF | MV leaflet description | Tethering | Radiographs | Notes |
| 1 | ME | 11.5 | 4 | 4 |  | Yes | Thickened leaflet tips, tenting, elongated anterior leaflet. | Posterior | Cardiomegaly (VHS 12), LAE, interstitial lung pattern | Treated with furosemide for one day prior to investigations |
| 2 | MN | 16.9 | 18 | 5 | developed AF 5 months later | Yes | Both thickened, poorly mobile | Both | Cardiomegaly (VHS 13.2), LAE, interstitial pattern, pulmonary venous congestion |  |
| 3 | MN | 19.3 | 54 | 3 | developed AF 4 months later | Yes | Tenting, thickened leaflets | Anterior | Cardiomegaly (VHS 12.7), LAE, mild interstitial lung pattern | Treated with furosemide and benazepril for one day prior to investigations |
| 4 | ME | 14 | 8 | 5 |  | Yes | Tenting, hockey stick appearance, elongated anterior leaflet | Posterior | Cardiomegaly (VHS 13.7), LAE, interstitial-alveolar lung pattern, pulmonary venous congestion |  |
| 5 | ME | 20.4 | 37 | 5 |  | Yes | Tenting, hockey stick appearance, thickened leaflet tips | No | Cardiomegaly (VHS 13.6), interstitial lung pattern, pulmonary venous congestion | PDA. CHF resolved and meds discontinued after ACDO |
| 6 | FN | 15.1 | 54 | 3 |  | No | Tenting, hockey stick appearance, elongated anterior leaflet | Posterior | Cardiomegaly (VHS 11), no lung pattern or venous congestion |  |
| 7 | MN | 16.5 | 50 | 3 |  | Yes | Hockey-stick appearance, tenting | Posterior | Cardiomegaly (VHS 14), no venous congestion or edema | On furosemide, pimobendan, benazepril and spironolactone for 5 days prior to presentation |
| 8 | FE | 11.4 | 15 | 3 | VPCs | Yes | Tenting, thickened leaflets, elongated anterior leaflet | Posterior | Cardiomegaly (VHS 15), mild pulmonary venous congestion, perihilar edema |  |

Supplementary table: Signalment and clinical information of the patients in the study. ACDO: Amplatz canine duct occluder, AF: atrial fibrillation, CHF: congestive heart failure, FE: female entire, FN: female neutered, LAE: left atrial enlargement ME: male entire, MN: male neutered, PDA: patent ductus arteriosus, VHS: vertebral heart score, VPC: ventricular premature complex

Figure titles and description

Figure 1. Echocardiographic images showing various mitral dysplasia lesions. Left apical 4 chamber views (A, C) and right parasternal views (B, D): A: thickened anterior leaflet, tethering of posterior leaflet to papillary muscles (end-systole); B: ‘hockey-stick’ appearance to valve leaflets (end diastole); C: elongated anterior leaflet, tethering of posterior leaflet to free wall (end diastole); D: elongated anterior leaflet and valve tenting (end systole)

Figure 2. Echocardiographic images displaying tenting of the mitral valve leaflets in early systole as seen from (A, C) a right parasternal long axis four chamber and (B, D) left apical four chamber view. In images C and D the tenting area is indicated by the cross-hatched region.

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| --- | --- | --- |
| Video 1 | 18 month male | Right parasternal long axis echocardiogram displaying thickened mitral valve leaflets and a ‘hockey stick’ appearance during diastole. |
| Video 2 | 50 month male | Left apical four chamber echocardiogram displaying thickened mitral valve leaflet tips with a ‘hockey stick’ appearance during diastole. Systolic tenting is evident. |
| Video 3 | 54 month female | Right parasternal long axis echocardiogram from the dog originally diagnosed with degenerative mitral valve disease. The mitral valve leaflet tips appear thickened, and the anterior leaflet is elongated. There is no valve prolapse and given the patient’s age, mitral dysplasia was suspected. |
| Video 4 | 15 month female | Right parasternal long axis echocardiogram showing an elongated anterior mitral valve leaflet and a short posterior leaflet that is tethered to the left ventricular free wall. There is a moderate eccentric jet of mitral regurgitation. Valve tenting is apparent in systole. |
| Video 5 | 8 month male | Right parasternal long axis echocardiogram from the dog originally diagnosed with dilated cardiomyopathy. The posterior leaflet and chordae are short and tethered to the papillary muscle; its movement is restricted. The anterior leaflet is elongated subjectively, and its diastolic movement also appears restricted, as the dog is not excessively tachycardic. Concurrent dilated cardiomyopathy is also possible, however the presence of primary valve abnormalities and the subjectively preserved systolic function would support a diagnosis of MD ~~with likely secondary myocardial failure~~ |
| Video 6 | 8 month male | Left apical four chamber echocardiogram from the dog originally diagnosed with dilated cardiomyopathy. The mitral valve leaflets have a slight ‘hockey stick’ appearance (diastolic doming) and display tenting in systole. |

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Footnotes

a Tristan Veterinary Practice Management Solution, version 1.8.3.1110

b GE Echopac version 113, GE Medical Systems, Buckinghamshire, UK

c Excel 2013, Microsoft Inc

d SPSS Statistics Version 24, IBM

e Van Israel, N., Dukes-McEwan, J., Biourge, V., Simpson, J.W.S. (2005). Athletic heart or DCM in a springer spaniel family? Proceedings of the 15th ECVIM-CA Congress. Glasgow, Scotland. 1-3 September 2005. Abstract 8. p. 193

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