CONGENITAL TRICUSPID VALVE MALFORMATION WITH EBSTEIN'S ANOMALY IN A GOLDEN RETRIEVER

Aangeboren dysplasie van de tricuspidalisklep met ebsteinanomalie bij een Golden Retriever

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ABSTRACT

A case of Ebstein's anomaly leading to right-sided heart failure in a one-year-old female Golden Retriever is reported. Clinical examination and investigations such as blood analysis, ECG, X-Ray, echocardiography and conservative therapy are described. Finally, the post mortem examination of the heart clearly showed the distal displacement of the malformed tricuspid valve leaflets, chordae tendinae and papillary muscles in the right ventricle, which is characteristic of Ebstein's anomaly.

SAMENVATTING

In deze casuïstiek wordt een anomalie van Ebstein beschreven die rechterhartfalen veroorzaakte bij een éénjarige Golden Retriever. Naast het klinisch onderzoek worden ook het bloedonderzoek, het EKG, de radiografieën, de echocardiografie en de mogelijke therapie besproken. Het post mortem onderzoek van het hart toonde een duidelijke distale verplaatsing aan van de misvormde tricuspidaalkleppen, chordae tendinae en papillair spieren in de rechterventrikel. Dit geheel van afwijkingen wordt beschreven als de anomalie van Ebstein.

INTRODUCTION

Tricuspid valve dysplasia (TVD) is defined as an uncommon congenital malformation of the whole tricuspid apparatus, including the tricuspid valve leaflets, the chordae tendinae and the papillary muscles, which usually results in tricuspid regurgitation (Kittleson and Kienle, 1998). Tricuspid valve dysplasia has been shown to have a genetic basis in the most commonly afflicted breed, Labrador Retrievers (Oyama and Sisson, 2001). One specific form of TVD, in which the basal attachments of the tricuspid valve are displaced ventrally into the right ventricle, is called Ebstein's anomaly (Kittleson and Kienle, 1998).

CASE HISTORY

A one-year-old female Golden Retriever weighing 20 kg became anorexic two days before presentation. Abdominal enlargement was present. The dog was small for her breed, but had always been very playful. The findings upon clinical examination, which are presented in Table 1, were suggestive of right heart failure. The dog was admitted to the hospital for further investigation.

At presentation, abdominocentesis was performed and furosemide (Lasix® Hoechst) was given IV 2 mg/kg and repeated one hour later. Analysis of the clear ascitic fluid (5 liters) revealed a modified transudate, characterized by a specific gravity of 1025, a total protein of 3.4 g/l and

Table 1. Clinical findings in the patient.

Clinical examination of	Findings
Body condition	Stunted growth
	Atrophy of temporal and longissimus dorsi muscles
Temperature	38.5° c
Mucous membranes	Pink; capillary refill time = two seconds
Jugular veins	Jugular venous distention without pulsation
	Positive hepatojugular reflux test
Femoral arterial pulses	Weak, without deficits
Respiratory rate	Tachypnea, 42 breaths per minute
Auscultation heart	100 bpm, regular rhythm
	Grade 3/6 systolic heart murmur with a pmi over the left apex
Lungs	No crackles
Abdomen	Ascites
Extremities	Warm

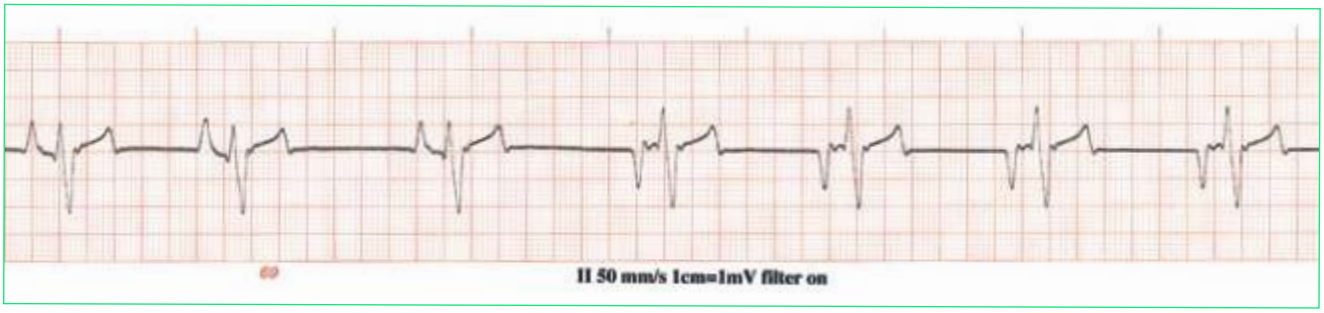


Figure 1. Lead II ECG indicating a sinus arrhythmia at 84 bpm with a wandering pacemaker and a bundle branch block pattern.

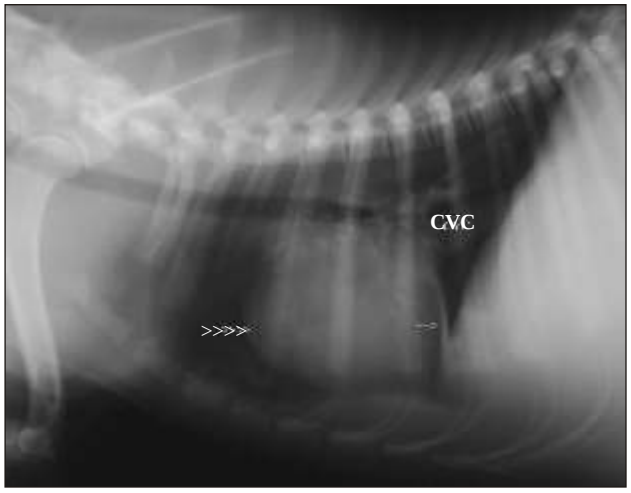
a cellularity above 2500 cells/mm^3 consisting of red blood cells and minor white blood cells with some mesothelial cells. The most relevant abnormality within the blood analysis was hypoproteinemia (47 g/l ref 57-77 g/l) due to protein loss in the ascetic fluid. Congestion of the liver was responsible for the slight hypoglycemia (3.4 mmol/l ref 3.5 – 6 mmol/l) and mild elevation of the alanine aminotransferase (49 U/l ref 15-30 U/L).

Electrocardiography revealed sinus arrhythmia (84 bpm) with a wandering pacemaker and a bundle branch block pattern (Figure 1).

Thoracic radiography showed right atrial and ventricular enlargement, with caudal vena cava engorgement and pleural fissures. Moreover, a clear heart apex shift to the left due to right heart enlargement could be demonstrated (Figure 2).

Echocardiography showed malformation of the tricuspid valve leaflets, chordae and papillary muscles, accompanied by an eccentric hypertrophy of the right ventricle and an apical displacement and malformation of the whole tricuspid annulus. Color flow Doppler showed a systolic insufficiency jet originating at the tricuspid valve and mitral valve (Figure 3A-D).

The dog was discharged on furosemide (Lasix®) 2 mg/kg PO q 12 hrs, spironolactone (Aldactone®) 2 mg/kg PO q 24 hrs and benazepril (Fortekor®) 0.5 mg/kg PO q



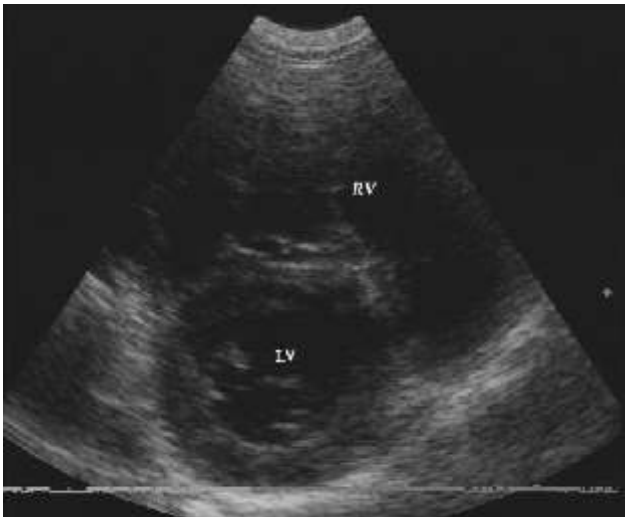


Figure 3A. Echocardiography - 2D right parasternal short-axis view shows a normal looking left ventricle (LV) and a large right ventricular outflow tract.

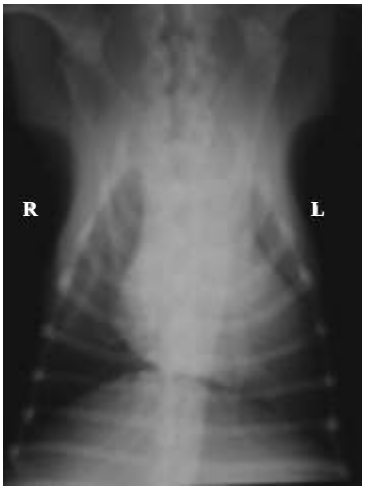


Figure 2. The left lateral radiography shows a right atrial and ventricular enlargement >>>> with caudal vena cava (CVC) engorgement and pleural fissures . The right radiography shows a clear heart apex shift to the left due to right heart enlargement.

	patient data	reference values*
RVDd	11.2	
IVSd	10.8	8.7 mm
LVDd	**27.6**	32.9 mm
PWd	10.4	7 mm
IVSs	9.2	13.1 mm
LVDs	20.8	19.9 mm
PWs	9.6	11.4 mm
FS	25	33-45%

Figure 3B. Echocardiographic measurements. M-mode 2D right parasternal short axis view, showing a large diameter of the right ventricle in diastole (RVDd) and a diminished diameter of the left ventricle in diastole (LVDd).
- VSd: interventricular septum in diastole
· PWd: free wall left ventricle in diastole
- IVSs: interventricular septum in systole
· PWS: free wall left ventricle in systole
- FS: fractional shortening
*** Boon, J, A. supplement.**

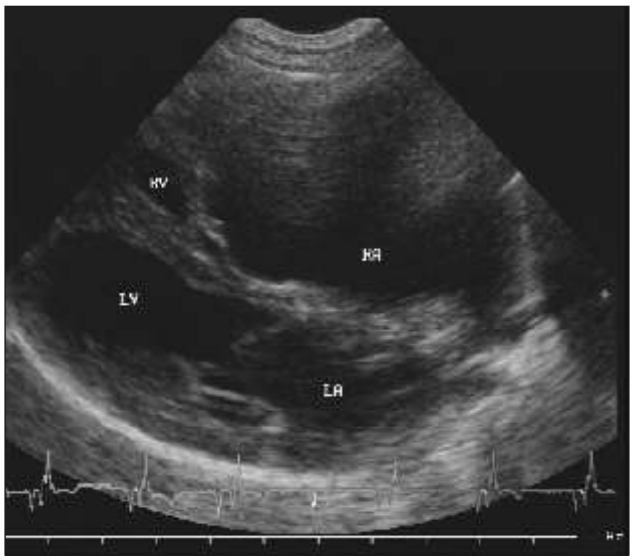


Figure 3C. Echocardiography - Right parasternal long-axis view showing apical displacement of the tricuspid valve, atrialization of the right ventricle (RV) and a huge right atrium (RA).

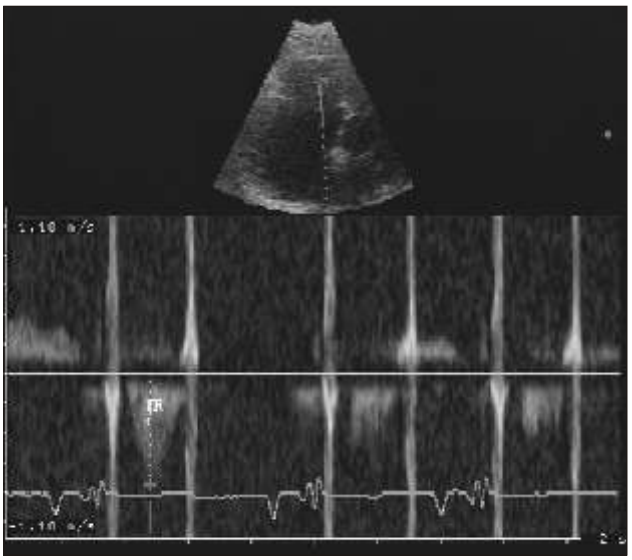


Figure 3D. Echocardiography - Left apical parasternal four-chamber view with tricuspid inflow. Continuous wave Doppler shows regurgitation of the tricuspid valve (TR).

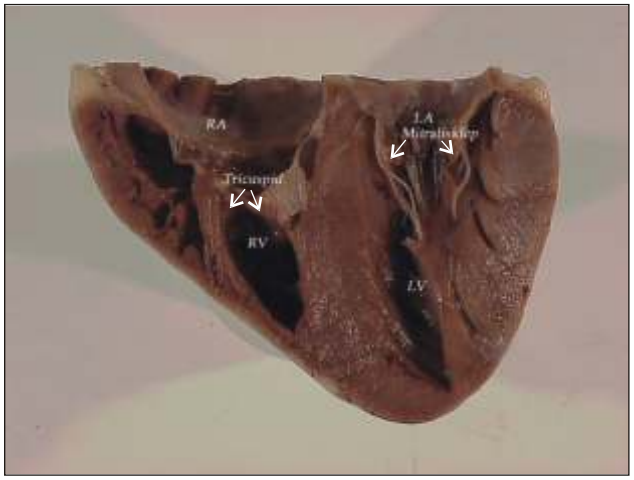


Figure 4. Showing tricuspid dysplasia and displacement with malformation of the valve leaflets. Marked enlargement of the right atrium. Left ventricle: mitral valve endocardiosis. RA= right atrium, RV= right ventricle, LA= left atrium, LV= left ventricle.

24 hrs. Two months after initial presentation, the dog was presented again with recurrent ascites. Due to the poor long-term prognosis, it was euthanized. Postmortem examination of the heart revealed severe right atrial and ventricular dilatation, thickened papillary muscle attachment of the mural leaflet of the tricuspid valve, septal leaflet of the tricuspid valve tethered to the interventricular septum by short chordae, distal displacement of the tricuspid annulus and slightly thickened and shortened mitral valves (Figure 4).

DISCUSSION

Tricuspid valve dysplasia is a congenital heart disease characterized by faulty development of the right atrioventricular valve leaflets, the chordae tendineae and the papillary muscles (Netter, 1981). An Ebstein's anomaly is a form of TVD characterized by a downward displacement of the leaflets so that "atrialization" of the right ventricle occurs (Moïse, 1989). The stunted growth of this dog, combined with its poor body condition was suggestive of a congenital defect.

At presentation, the dog showed distended jugular veins, ascites, weak femoral pulses, hepatomegaly with a positive hepatojugular reflux and pleural effusions, all indicative of right heart failure. Clinically, TVD is characterized by a holosystolic or pansystolic right apex murmur (Kittleson and Kienle 1998). In this case, no right-sided murmur was detected. This was probably due to severe dilation and malformation of the tricuspid annulus, producing a laminar regurgitant flow. The left side murmur in this case was due to concurrent mild mitral dysplasia, which was seen on pathology. In many cases of TVD, the mitral valve is also affected (Fox *et al.*, 1999).

The analysis of the ECG showed an increase in P-wave duration and amplitude, which is indicative of biatrial enlargement. The changing configuration of the P-waves could be due to wandering pacemaker, but is in this case more suggestive of intra-atrial or interatrial conduction disturbances (Tilley, 1992). In the present case, the right axis deviation of -120°, the S-waves in the standard leads I, II, III and aVF, and the S-waves larger than 0.35 mV in lead II point to right heart enlargement (Tilley, 1992). The splintered QRS-complexes are distinctive and common electrocardiographic findings in dogs with TVD (Moïse, 1995). The notched QRS-complexes, as seen in this case, are more common than the Rr', RR', rR' and rr' morphologies (Konreich and Moïse, 1997). The cause of these splintered QRS patterns is unknown, but suggested mechanisms for this altered ventricular depolarization pattern include fibrosis, bypass tracts, or ischemia (Moïse, 1995). Microscopic intramural myocardial infarction can also be responsible for notched QRS complexes, which are more often seen in elderly dogs. The intraventricular conduction defect was represented by a right bundle branch pattern. The repolarization of these ventricles caused an enlarged T-wave.

Thoracic radiography revealed a severely dilated right atrium, causing a large bulge on the cranial aspect of the heart, and a left apex shift on the dorsoventral view. The caudal vena cava-to-aorta ratio of 1.5 was suggestive of right-sided heart failure (Thrall, 2002). In this case, engorgement of the caudal vena cava, hepatomegaly, pleural effusion and ascites indicated right heart failure.

Echocardiography showed the right atrium to be the largest of all four chambers. The right ventricle was very small due to the apical implantation of the tricuspid valve. With TVD, the left heart looks small, which is expressed by reduced end-diastolic and end-systolic diameters (Kittleson and Kienle 1998). In this case, only the end-diastolic diameter of the left ventricle is not likely to be underloaded as expected, because of the concurrent mitral valve dysplasia, which might be responsible for a left ventricle eccentric hypertrophy. The increased size of the hepatic veins on echographic examination supported the diagnosis of right heart failure.

The treatment objectives of chronic right-sided congestive heart failure include control of the effusions and stopping of the neuro-endocrine cascade associated with heart failure. Initial stabilization in this patient was accomplished by abdominocentesis and intravenous administration of furosemide. Chronic therapy of right-sided heart failure due to tricuspid regurgitation includes diuretics, ACE Inhibitors and digoxin (Fox *et al.*, 1999). In this case, the loop diuretic furosemide combined with the

potasium-sparing diuretic spironolactone were administered to further relieve the effusions. Benazepril is used mainly to stop the neuro-endocrine cascade, but also to improve pump function by reducing afterload (right ventricle) and to counteract myocardial remodeling. Repeated abdominocentesis is often required when ascites is refractory to drug therapy (Fox *et al.*, 1999), but this may result in marked hypoproteinemia and should be monitored carefully. Refractory ascites was the reason for euthanasia in this case.

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