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# Tricuspid valve dysplasia complicated with pulmonic regurgitation in a Cocker Spaniel dog

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**Abstract :** A 17-month-old intact male Cocker Spaniel was presented with primary complaints of severe ascites, exercise intolerance, and diarrhea. Diagnostic studies revealed tricuspid and pulmonic regurgitation on phonocardiogram, right ventricular enlargement on the electrocardiogram, typical right cardiac enlargement signs on the thoracic radiography and tricuspid valve malformation and marked enlargement of the right atrium and right ventricle on the echocardiography and tricuspid and pulmonary regurgitation on the color spectral echocardiography, suggesting tricuspid valve dysplasia and pulmonary hypertension. Using angiography and cardiac catheterization, pulmonary hypertension was ruled out. Further echocardiographic study revealed membranous valvular structures cranial to pulmonary annulus causing pulmonary regurgitation. Based on these findings on the diagnostic investigation, the case was diagnosed as tricuspid valve dysplasia complicated with pulmonic regurgitation. The dog was medically managed with furosemide, enalapril, nitroglycerine transdermal patch and pimobendan after the ascitic fluid removal.

**Keywords :** AV valvular malformation, dog, tricuspid valve dysplasia

## Introduction

Tricuspid valve dysplasia (TVD) is a congenital heart defect in tricuspid valvular structures including valvular leaflets, chordae tendineae, and/or papillary muscles and causes backflow from right ventricle (RV) to right atrium (RA; valvular regurgitation) due to incomplete closure of tricuspid valve [5]. If the cellular degeneration to separate the flaps of the tricuspid valve from RV wall which normally occurred during the embryonic development fails to take place, this results in regurgitation of blood back into the RA due to the valvular insufficiency. Due to the backflow from the RV, the RA expands to accommodate the backflow from the RV. Consequently, the development of eccentric left ventricle (LV) hypertrophy from volume dilation of RA will cause the increase in size of tricuspid valve annulus leading to worsened tricuspid valve regurgitation. Right-sided congestive heart failure is the end-result and is characterized by ascites, hepatomegaly and/or jugular vein distension.

Wide spectrum of abnormalities in tricuspid valvular structures can cause TVD but almost always result in tricuspid valve regurgitation. According to human literature, TVD can be occurred by i) focal or diffuse thickening of the valve leaflets, ii) underdevelopment of chordae tendineae and papillary muscles, iii) incomplete separation of valve components from the ventricular wall, iv) focal agenesis of valvular tissue and v) any combination of this abnormalities [8]. Most common abnormality in dogs and cats with TVD is the underdevelopment of chordae tendineae, which causes the direct attachment of papillary muscles to the valve leaflets [6]. Ebstein's anomaly is an extreme case of TVD and is characterized by the downward displacement of septal leaflet and atrialized right ventricle [1, 9].

Although TVD is a rare congenital heart defect, it has been reported in numerous dog breeds including old English sheepdogs, great Danes, German shepherds, and Irish setters. Recent study also found a preponderance of Labrador retrievers as an inherited genetic disorder although the clear inheritance mode

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has yet been defined [2].

In this case report, we described a rare case of tricuspid valve dysplasia complicated with pulmonic regurgitation in a Cocker Spaniel dog.

## Case presentation

A 17-month-old intact male Cocker Spaniel was presented to the Veterinary Teaching Hospital, Kangwon National University with signs of severe ascites, exercise intolerance, and diarrhea. On thoracic auscultation, a grade IV / VI systolic regurgitation murmur and systolic-diastolic regurgitation murmur were heard over the right apex and the left base, respectively (Fig. 1).

On the day of presentation, electrocardiographic (ECG) studies showed splintered QRS complexes and the presence of S-waves in the frontal leads indicating right ventricular hypertrophy (Fig. 2A) and splintered QRS complexes (RSr') characteristic of tricuspid valve dysplasia in precordial leads (Fig. 2B). Prominent R waves on V1 right chest lead and the presence of S waves in V3-V5 left chest lead were are also suggested right ventricular hypertrophy (Fig. 2B).

Routine hematology and biochemistry showed no particular abnormalities except hypoproteinemia (total protein 4.6 g/dl, reference range 5.3-7.6 g/dl, albumin 2.2 g/dl, reference range 3.1-4.7 g/dl). Abdominal fluid analysis revealed hemorrhagic modified transudate (protein 3 g/dl, ~5,000 cells/ul).

Radiographic studies of the thoracic and abdominal cavities revealed a globoid cardiac shadow, increased

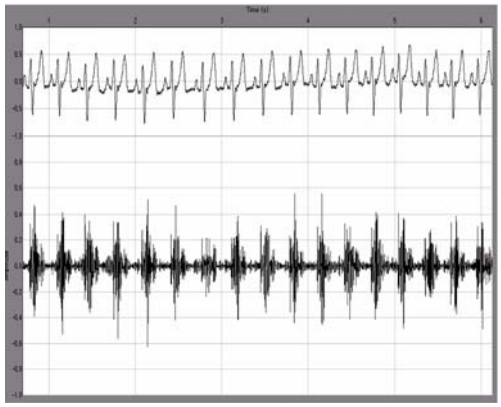

**Fig. 1.** Phonocardiogram of this case recorded at the right apex. There are decrescendo quality murmurs at systole (tricuspid regurgitation) and diastole (pulmonic regurgitation).

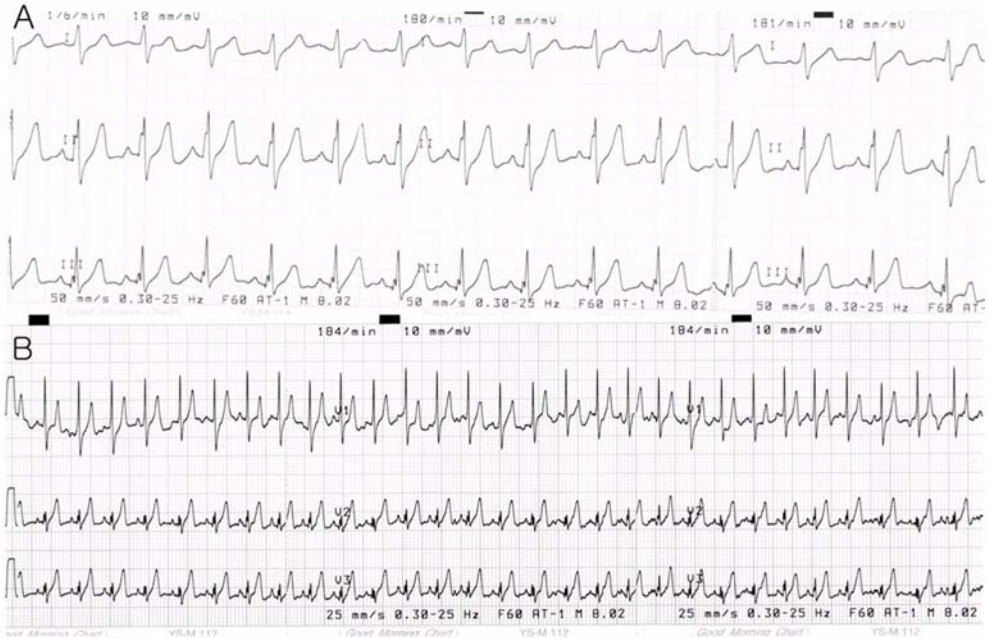


**Fig. 2.** Electrocardiogram (ECG). A: The frontal leads of ECG showed splintered QRS complexes and the presence of S-waves B: the precordial leads showed splintered QRS complexes (RSr'), prominent R waves on V1 right chest lead and the presence of S waves in V3 left chest lead.

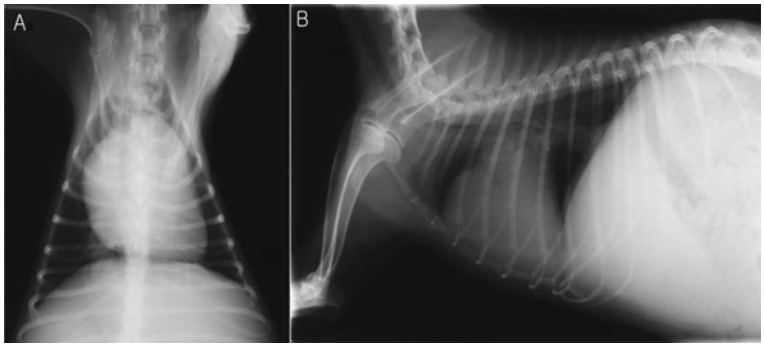


**Fig. 3.** Thoracic radiography A: Dorsoventral projection of the radiography showed marked cardiomegaly with right ventricular enlargement. B: Lateral projection of the radiography showed increased cardiac sternal contact, a distension of the caudal vena cava, caudal gastric axis deviation, an enlarged hepatic shadow, and ascites. Marked dilation of the root of main pulmonary artery was also remarkable on this view.

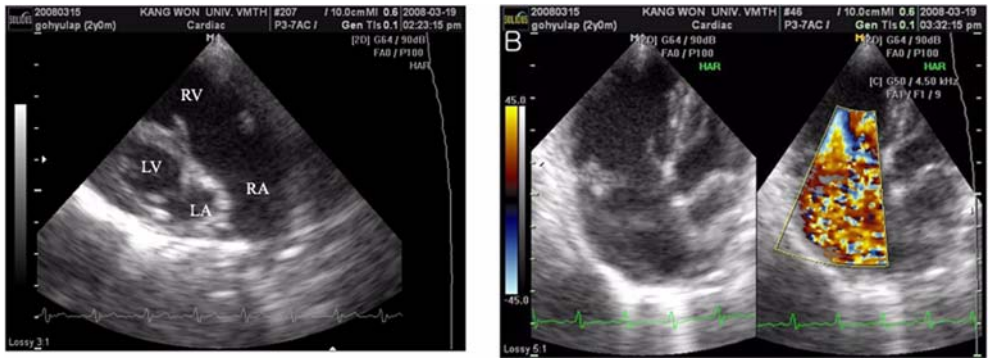


**Fig. 4.** The echocardiography of this case. A: On this echocardiogram, the septal leaflet of tricuspid valve (TV) appeared to be adhered to the interventricular septum, whereas the mural leaflet of TV was also enlarged and thickened. The dimensions of right atrium (RA) and ventricle (RV) were markedly enlarged, while those of left atrium (LA) and ventricle (LV) were relatively reduced. B: Echocardiogram showed paradoxical movement of tricuspid valvular leaflets (unharmonious valvular coaptation with mural leaflet; left) and severe regurgitant flow from RV to RA in systole.

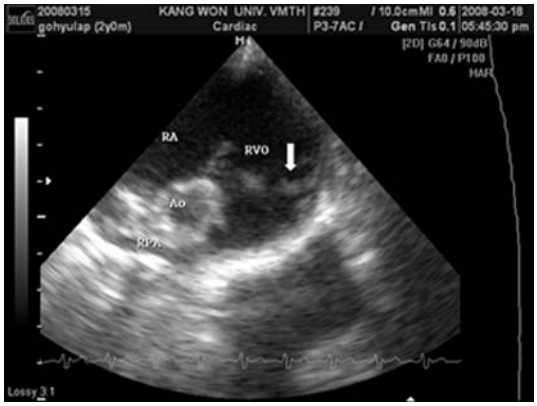


**Fig. 5.** The echocardiography of this case at pulmonic valvular area. Echocardiography showed abnormal membranous valvular structures (arrow) cranial to the actual pulmonic valvular annulus causing regurgitant turbulent flow.

cardiac sternal contact, a distension of the caudal vena cava, caudal gastric axis deviation, an enlarged hepatic shadow, and ascites, suggesting right-sided congestive heart failure (Fig. 3). Marked dilation of the root of main pulmonary artery (MPA) was also obvious in lateral thoracic radiography (Fig. 3B).

A 2-dimensional echocardiographic examination found that the septal leaflet of tricuspid valve (TV) appeared to be adhered to the interventricular septum (Fig. 4A) and have paradoxical movement (unharmonious valvular coaptation with mural leaflet, Fig. 4B left). Further examination also found that marked enlarged RA and severe regurgitant flow from RV in systole (Fig. 4B) and relatively reduced dimension of left atrium (LA) and LV (Fig. 4A). The mural leaflet of TV was also enlarged and thickened (Fig. 4A).

Echocardiography at pulmonic valvular area revealed abnormal membranous valvular structure cranial to the actual pulmonic valvular annulus causing regurgitant turbulent flow (Fig. 5). Color Spectral Doppler study revealed moderate tricuspid regurgitant (TR) flow with a peak velocity of 2.4 m/s (pressure gradient between RV to RA; 23 mmHg; Fig. 6A) and pulmonic regurgitant (PR) flow with a peak velocity of 1.1 m/s (pressure gradient between pulmonic artery (PA) to RV; 5 mmHg; Fig. 6B). The M-mode echocardiography at left ventricular papillary muscle level revealed marked reduction of fractional shortening (%FS; 14.38%), left ventricular ejection fraction (LVEF; 33.51%), flattening of interventricular septum and marked right ventricular dilation (Fig. 7A).

Based on our diagnostic findings including tricuspid and pulmonic regurgitation on phonocardiogram, right ventricular enlargement on ECG, typical right cardiac enlargement signs on the thoracic radiography and tricuspid valvular malformation and marked enlargement of RA and RV on echocardiography, possible diseases are tricuspid valvular dysplasia, Ebstein's anomaly, pulmonic stenosis, pulmonic hypertension, pulmonic valvular dysplasia or in combination of those anomalies.

Because our radiography showed marked dilation of root of MPA (although no stenotic jet flow observed on the echocardiography), transvenous angiography was performed to rule out pulmonic stenosis and pulmonic hypertension (PHT; Fig. 8A). However angiographic studies failed to find stenotic pulmonic

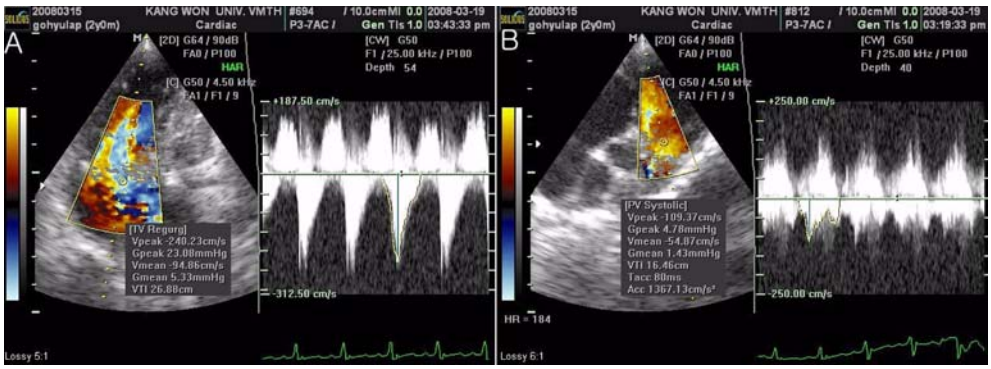


**Fig. 6.** Color spectral Doppler echocardiography of this case. A: Echocardiogram taken at tricuspid valvular area moderate tricuspid turbulent regurgitant flow with a peak velocity of 2.4 m/sec (pressure gradient between right ventricle (RV) to right atrium; 23 mmHg) at systole. B: Echocardiogram taken at pulmonic valvular area mild pulmonary turbulent regurgitant flow with a peak velocity of 1.1 m/sec (pressure gradient between pulmonic artery to RV; 5 mmHg) at systole and diastole.

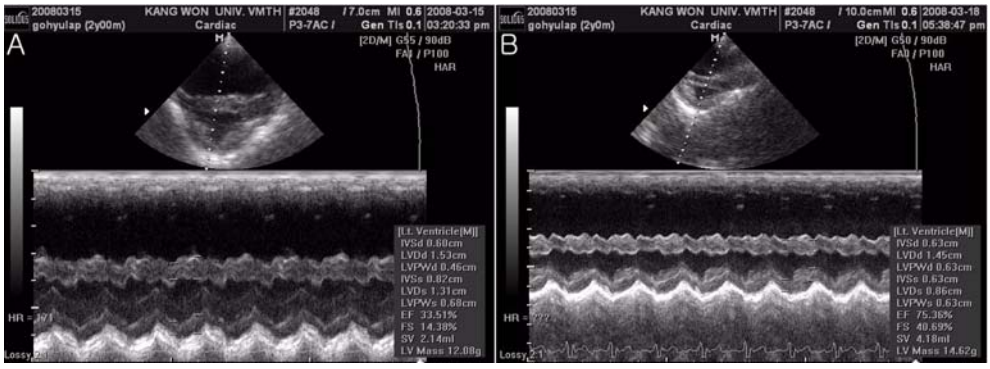


**Fig. 7.** The M-mode echocardiography of this case taken at left ventricular papillary level. A: Before the initiation of medical treatment. The M-mode echocardiography showed marked reduction of fractional shortening (%FS; 14.38%), left ventricular ejection fraction (LVEF), flattening of interventricular septum and marked right ventricular dilation. B: 3 days after the medical treatment including diuretics (furosemide and spironolactone), angiotensin converting enzyme inhibitors (enalapril), inotropics (pimobendan), and nitrate patch. The M-mode echocardiography showed marked improvement in %FS (40.69%) and LVEF, although right ventricular dimension was not much changed.

lesions. Furthermore there was no dilation of right or left pulmonary artery, which could exclude the possibility of pulmonic hypertension (Fig. 8A). One interesting finding on the angiography is the back flow from RA to jugular vein, when the contrast media injected into the RA, suggesting a marked increased RA pressure possibly due to the regurgitant flow from the RV (Fig. 8B). However, we were unsure how severe volume or pressure overload in the right sided cardiac chambers. Because TR and PR could be occurred by severe PHT, more definitive diagnostic test was required for the differentiation TVD from PHT. Therefore the blood pressure in RA, RV and PA were measured using cardiac catheterization with 5-Fr micromanometer-tipped catheter (Microtip catheter transducer SPC-350; Millar instruments, USA). The pressures in RA, RV and PA were 20-25/8-10 mmHg, 42-34/3-8 mmHg, 20-25/10-13 mmHg, respectively, indicating marked increased RA pressure in systole and diastole and RV pressure in systole, but normal pressure of PA in systole and diastole (Fig. 9). Those findings helped to rule out the PHT in our case. Based on our findings on the diagnostic investigation, the case was diagnosed as tricuspid valve dysplasia complicated with pulmonic regurgitation.

The dog was treated initially with furosemide (2 mg/kg, PO, BID; Daewon Pharmaceuticals, Korea) and enalapril (0.5 mg/kg, PO, BID; Daewon Pharmaceuticals, Korea), and fed a salt restricted diet (Hill's h/d; Hill's Pet Nutrition, USA) for alleviating the circulatory failure. In addition, 1 l of ascitic fluid was removed by the abdominocentesis. Although the clinical condition of the patient was dramatically improved, the ascites was recurred 2 days after the treatment. Therefore, the abdominal fluid was removed again with addition of venodilator therapy (nitroglycerin patch; 3.73 mg/head,

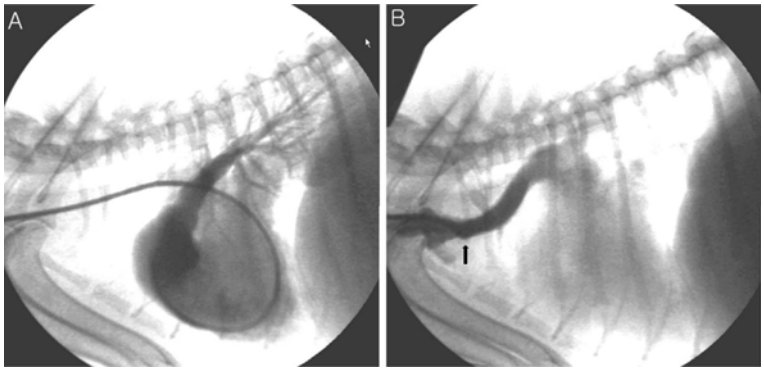


**Fig. 8.** Transvenous angiography of this case. A: There is marked dilation of the root of the main pulmonary artery. However, this angiography failed to find stenotic pulmonic lesions and dilation of right or left pulmonary arterial branches. B: There is a substantial amount of back flow from the right atrium (RA) to the jugular vein due to increased intracardiac chamber pressure in the RA. See the location of the catheter tip (arrow).

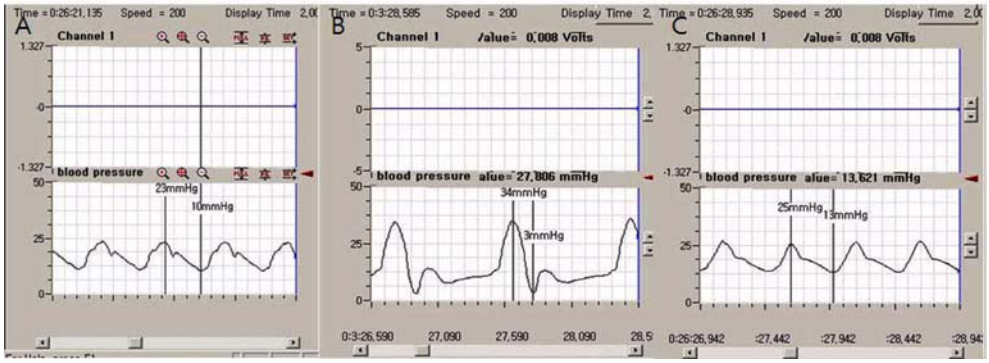


**Fig. 9.** The measurement of intracardiac chamber pressure of this case. The pressures in right atrium (A), right ventricle (B) and pulmonary artery (C) were 20-25/8-10 mmHg, 42-34/3-8 mmHg, 20-25/10-13 mmHg, respectively.

transdermal, every other 12 h, Angiderm patch; Samyang, Korea), spironolactone (1 mg/kg, BID, Aldactone; Pfizer, Korea) and an inotropic agent, pimobendan (0.25 mg/kg, PO, BID, Vetmedin; Boeringer Ingelheim, Switzerland). After changing medication, the abdominal fluid accumulation was subsided. The M-mode echocardiography taken 3 day after the medical treatment showed marked improvement in %FS (40.69%) and LVEF (75.36%), indicating the overall improvement in cardiac performance (Fig. 7B).

After 3 months of medical treatment, the clinical signs were worsen (e.g. reappearance of ascites). Therefore, the dosages of furosemide and nitroglycerine were escalated to 3.0 mg/kg and 5.6 mg/head, respectively. Although the clinical signs were improved, the ascites was re-occurred at 2-3 weeks intervals. Currently the furosemide is switched to torsemide (0.3 mg/kg, PO, BID; Ildong Pharm, Korea) to minimize azotemia and hypokalemia. The fluid accumulation is managed by periodical abdominocentesis. Echocardiographical studies taken after 8 months of medical treatment, overall cardiac performance was maintained well (%FS; 45.19% and LVEF; 79.27%) with minimal worsening of TR (2.2 m/sec) and PR (1.7 m/sec). The dog is currently being monitored monthly interval and maintained normal physical viability.

## Discussion

Diagnostic features of TVD are i) cardiomegaly with massive RA enlargement on the thoracic radiography [4], ii) right apical systolic murmur [5], iii) high P waves, deep S waves in leads I, II, III, and aVF, and splintered QRS complexes on the ECG [7], iv) markedly enlarged RA and RV, often smaller size LA and LV than normal, and abnormal sepal tricuspid leaflet adhered to the interventricular septum or to the papillary muscles [3] on the echocardiography, and a turbulent regurgitant flow into RA during systole on the color spectral Doppler studies [6]. The dog presented showed all features of TVD in our diagnostic investigation. However, the main cause of TR in dogs was tricuspid valvular insufficiency and PHT. Because the dog showed moderate TR and mild PR simultaneously in the initial diagnostic studies, we had to investigate further to rule out PHT using more invasive diagnostic procedures (e.g. angiography and catheterization). However, our invasive diagnostic studies clearly ruled out PS and PHT, because there was no narrowing on right ventricular outflow tract and no dilation on the right and/or left pulmonary arteries. Moreover, the pressure in pulmonary arteries (including wedge pressure) was not as high as seen in PHT in this case, eliminating the possibility of PHT. However, those diagnostic studies did not still clearly answered the presence of PR and marked dilation of PA root, since the pressure gradient between RV to PA was not as high as to cause PR, based on the intracardiac pressure measurement. However, our further echocardiography clearly revealed abnormal membranous valvular structures cranial to pulmonary annulus causing PR in this case. The turbulent at this lesion might cause the marked dilation of the PA root. Fortunately, these structures did not cause the obstruction of right ventricular outflow tract and obstruct the blood flow from RV to PA much.

Medical treatment for TVD is the best option at this moment, because of unfavorable outcome from surgical treatment. Therefore medical treatment is usually directed to improve quality of life by reducing the amount of abdominal fluid periodically and by medicating certain drugs to slow down the fluid accumulation. Diuretics and angiotensin converting enzyme inhibitors (ACEi) are the common drug for this purpose [6]. Such medical therapy is commonly effective at slowing down the time until the abdomen is severely distended again. However, the complete stopping of fluid accumulation is often not effective and necessary on the view of long-term management [6]. Therefore most TVD cases required periodic abdominocentesis and fluid removal. However, periodic removal of abdominal fluid causes substantial loss of plasma albumin, although it rarely results in significant complication. However, with time, because the interval for ascetic fluid removal will become too short, other medications (e.g. inotropes, venous dilators, spironolactone) may be helpful to slow down the fluid accumulation, as seen in this case. Due the severity of our case, the combination therapy using furosemide and ACEi was not enough for the retardation of fluid accumulation. After the addition of inotropes and venous dilators, we could delay the further accumulation of ascitic fluid. Therefore our case study suggested the addition of inotropes and venous dilators might be the good option for slowing fluid accumulation frequently seen in the right sided heart failure.

Although TVD in dogs has been already published in Korea [8], our case study described diagnostic and therapeutic strategies for TVD case complicated with PR by abnormal structures, which was difficult to differentiate from PHT without more invasive diagnostic studies. Therefore our case study may be valuable resource for identifying more complicated case of congenital heart defects in small animal practice.

## Acknowledgment

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