J Vet Clin 25(3) : 195-199 (2008)

Journal of Veterinary Clinics
www.ksvc.or.kr

Partial Atrioventricular Canal Defect in a Maltese Dog

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(Accepted: April 4, 2008)

Abstract : A 2-year-old female Maltese dog was referred with primary complaints of exercise intolerance and abnormal heart sound. Clinical and diagnostic investigation revealed split S2 and S4 gallop in the cardiac auscultation, tall P wave and left anterior fascicular block in the electrocardiogram, left atrial enlargement on the thoracic radiography, ostium primum atrial septal defect and cleft of the anterior leaflet of the mitral valve on the echocardiography. Based on those findings, the dog was diagnosed as the partial atrioventricular canal defect. Since the dog showed mild exercise intolerance, enalapril and furosemide were prescribed.

***Key words* :** partial atrioventricular canal defect, ostium primum atrioventricular septal defect, dog, S4 gallop, fascicular block.

Introduction

Atrioventricular canal defects (AVCDs) are caused by an abnormal or inadequate fusion of the superior and inferior endocardial cushions with the mid portion of the atrial septum and the muscular portion of the ventricular septum. AVCDs have also been called endocardial cushion defects, atrioventricular septal defects, and persistent atrioventricular ostium (7). AVCDs are classified into partial and complete atrioventricular canal defects, depending on morphology of atrioventricular defect.

Complete AVCDs have the three major abnormalities including a septum primum atrial septal defect, a high ventricular septal defect and one abnormally formed five-leaflet of atrioventricular valve (1,10), while partial AVCD is characterized by ostium primum atrial septal defect (ASD) and a cleft of the anterior mitral leaflet. Ostium primum ASD may exist in isolation however, most commonly coexist with a cleft in the anterior leaflet of the mitral valve (7,10). In this type of defect, the mitral and tricuspid annuli are separate. In addition, partial AVCD should have various anatomic abnormalities related with atrioventricular valve, atrial septum and left ventricular outflow tract (7,15).

Partial AVCD have the ostium primum ASD in which the large defects contribute no resistance to flow (8). In this situation, only the compliance of the two ventricles determines impedance of diastolic flow. Thus, in conjunction with diastolic time, it determines the amount of flow (7). The aetiologies of partial AVCD can be explained simply on the following factors including mutations in transcription factors associated with cardiomorphogenesis (e.g. NKX2.5, TBX5, and GATA4), trisomy 16, and various abnormal cardiac developmental elements (3-5). AVCDs are one of the most common congenital heart diseases in cats (9). However, partial AVCSs are rare in dogs and cats (10,11,13).

The following report documents a rare case of partial AVCDs in a Maltese dog. The aims of this case study are to provide clinical and diagnostic features of AVCDs in a dog and to enrich our resources for rare congenital heart defects.

Case Description

A 2-year-old female Maltese dog (3 kg of body weight) was referred to the Veterinary Teaching Hospital, Kangwon National University with complaint of gallop sound and mild exercise intolerance. On phonocardiogram, the S4 gallop and split S2 were detected over the left basal area (Fig. 1). Electrocardiogram revealed a left anterior fascicular block characterised by an upright QRS complex in lead I, negative QRS complex in aVF, and left axis deviation of the mean electrical axis (-41°). P-wave was positive in leads I, II and III, and the amplitude of the P wave in lead II was significantly increased (0.5 mV), suggesting right atrial enlargement (Fig. 2). No significant abnormalities were observed in complete blood count (CBC; hematocrit 45%, haemoglobin 16 g/dL) and blood biochemistry.

Thoracic radiography revealed no cardiomegaly (VHS 9.2, reference range: 8.5-10.5) with a mild left atrial enlargement on lateral projection (Fig. 3A). No particular abnormalities were observed on the ventrodorsal projection (Fig. 3B).

The 2-dimentional echocardiography showed right ventricular and biatrial enlargement, defect of septum primum (ostium primum), prolapsed septal leaflet of the tricuspid valve (Fig. 4A), and a cleft in the septal cusp of the mitral valve (Fig. 5). The interventricular septum was intact. On the

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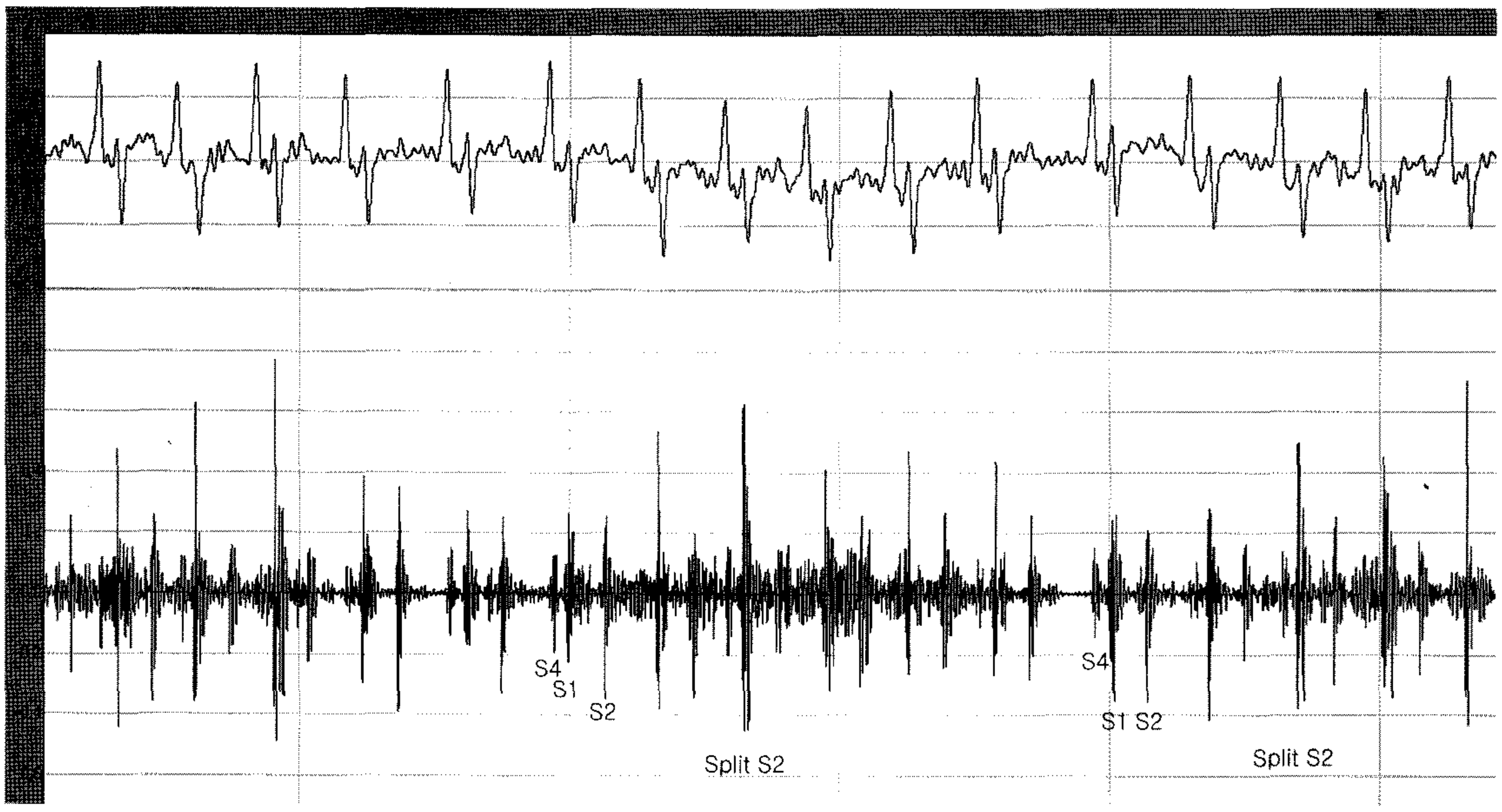

Fig. 1. Phonocardiogram of this case. Phonocardiography is clearly showing split S2 and S4 gallop.

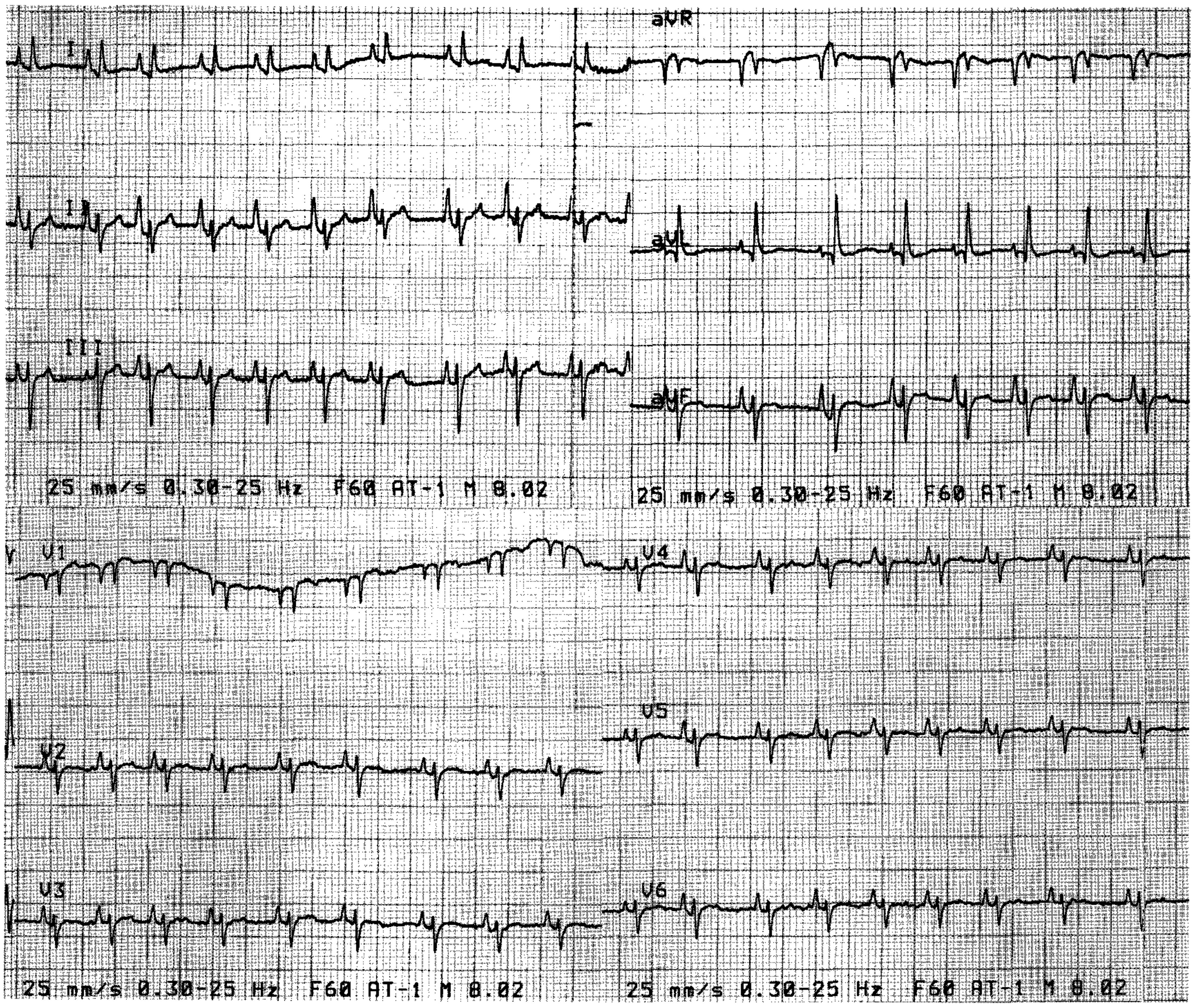

Fig. 2. The 12-lead electrocardiogram recorded from this case. The heart rate is approximately 165 beats/min. The ECG tracing showed a characteristic left anterior fascicular block (Positive QRS complexes in lead I, negative QRS complexes in lead aVF, and left axis deviation; -41°) and tall P wave (0.5 mV; reference range, maximum 0.4 mV).

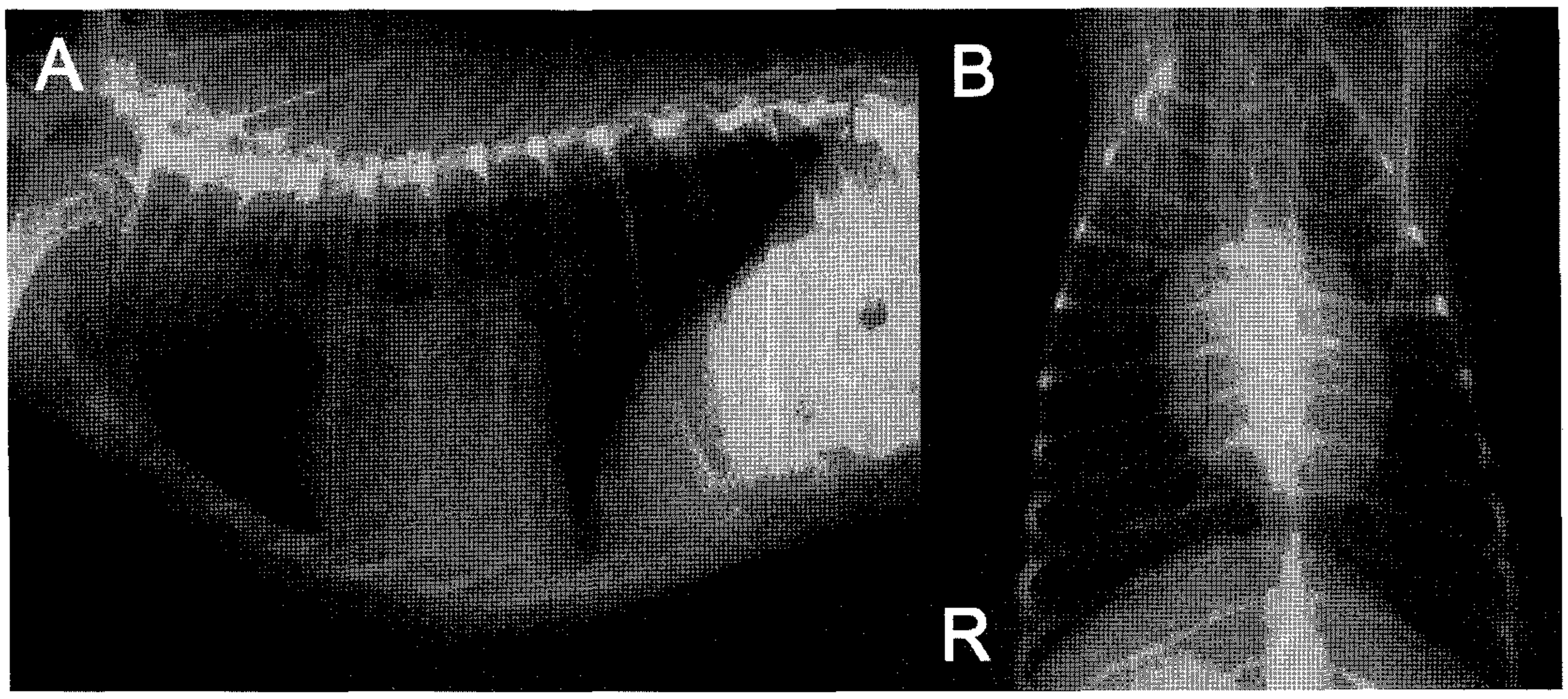

Fig. 3. Thoracic radiography of this case. A: Right lateral projection: Thoracic radiograms revealed no cardiomegaly (VHS 9.2, reference range: 8.5-10.5) with a mild left atrial enlargement. B: Dorsoventral projection: No particular abnormalities were observed.

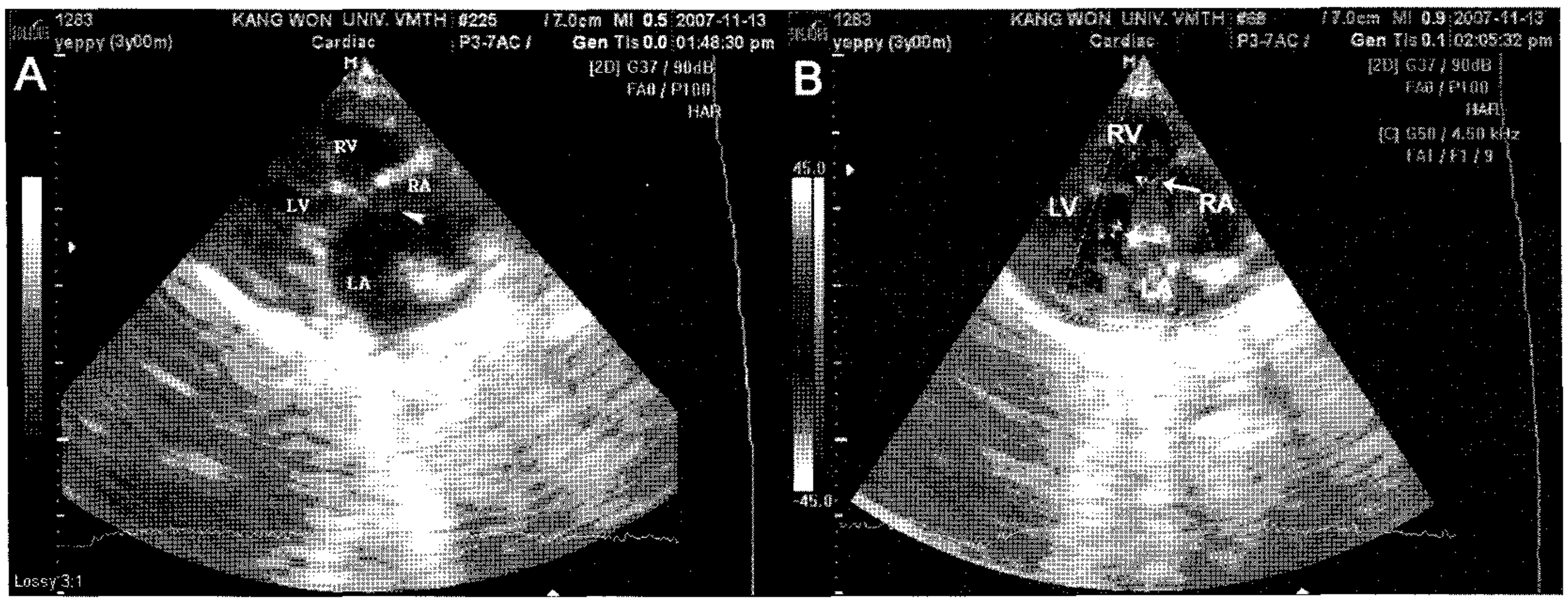

Fig. 4. Two-dimensional and colour Doppler echocardiography of this case. The view is a right parasternal long-axis view. A: The 2-dimentional echocardiography showed right ventricular and biatrial enlargement, defect of septum primum (ostium primum), prolapsed septal leaflet of the tricuspid valve (white arrow head). B: Colour Doppler echocardiography showed mild turbulent flow at the tricuspid valve and mitral valve region.

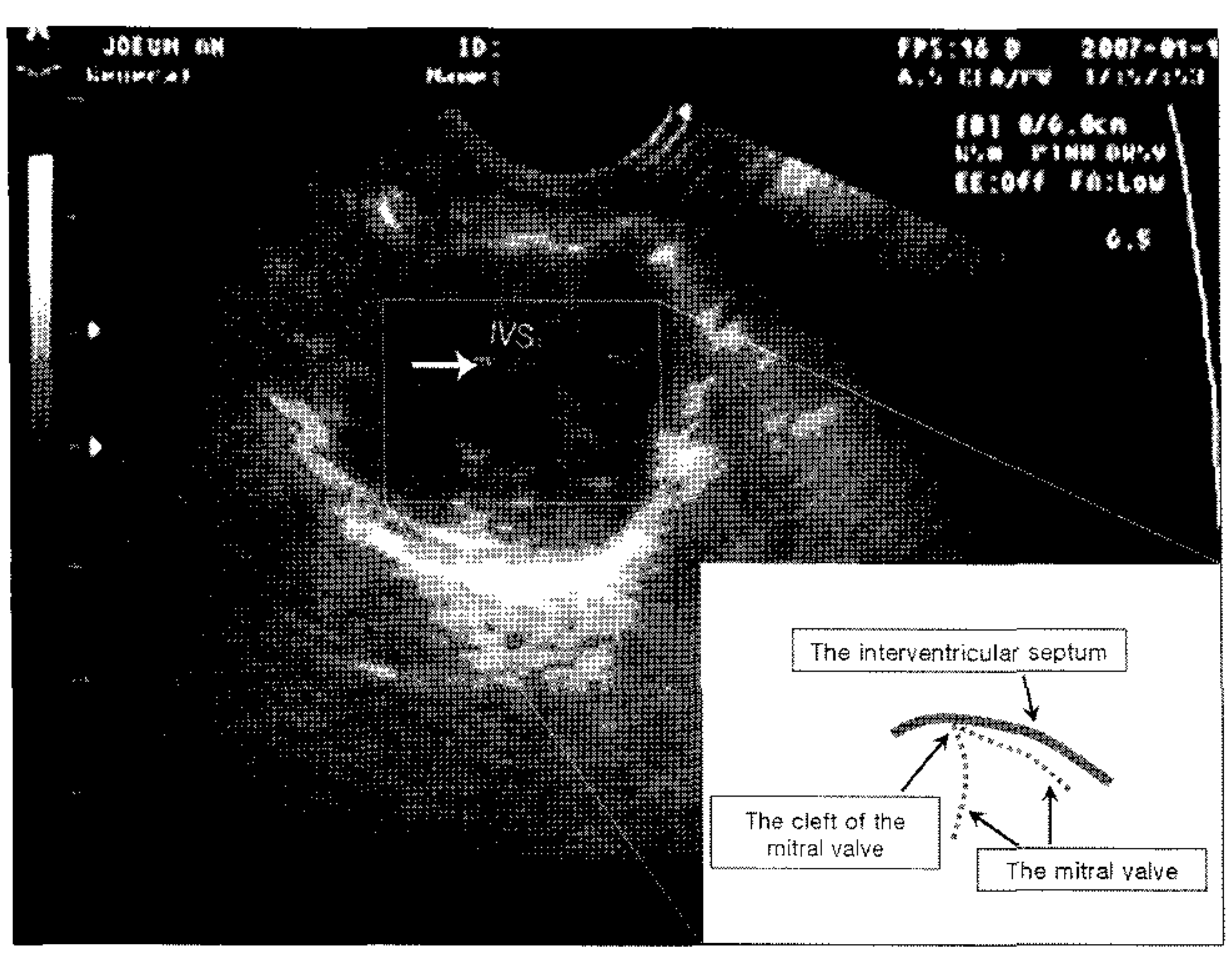

Fig. 5. The right parasternal short-axis view of this case. A cleft (white arrow) in the septal cusp of the mitral valve is visualized at the level of the mitral valve. The right bottom of this figure is schematic drawing of this figure.

right parasternal long-axis four-chamber view of colour-flow Doppler echocardiography, left to right shunt across the septal defect was evident. Furthermore, on this view, the mild turbulent flow at the tricuspid valve and mitral valve region was detected, indicating atrioventricular valve insufficiency and regurgitation (Fig. 4B). Based on these findings, the case was diagnosed as partial atrioventricular canal defects complicated with left anterior fascicular block.

Since the dog showed mild exercise intolerance, this patient was prescribed with angiotensin converting enzyme inhibitor (ACEi; Enalapril, 0.5 mg/kg, bid, PO) and diuretic (furosemide, 2 mg/kg, bid, PO). To date, the patient is well managed with medical treatment. The patient is being monitored with a routine medical check-up at regular intervals.

Discussion

Partial AVCD is characterized by ostium primum atrial septal defect (ASD) and a cleft of the anterior mitral leaflet and is easily misdiagnosed as isolated ostium primum ASD (10). The isolated ostium primum ASD is the simplest form of AVCDs (1). Pathogenesis of the partial AVCD is similar with ostium primum ASD. However, since the mitral and tricuspid annuli are separate, the treatment of the partial AVCD includes the surgical correction of the cleft of the AV valve beside ostium septum defect (10,11)

The common abnormal heart sound of the partial AVCDs is systolic ejection murmur at the left base and split S2 sound (1,7). The regurgitant murmur may be detected at the left and right heart apex, depending on the size of cleft of the atrioventricular valves (13). On the phonocardiogram, this patient showed split S2 and S4 gallop, which were characteristic abnormal heart sound associated with AVCDs (1,7). The split S2 of our case might be caused by prolonged right ventricular ejection time longer than left ventricular ejection time, due to shunt flows from the left atrium causing right ventricular volume overload. Therefore, the right ventricular ejection time of this dog was prolonged because the amount of blood in the right ventricle was markedly increased, which was well described in veterinary literatures (1,7). Furthermore, the large defect such as ostium primum ASD of this case might result in eccentric hypertrophy of the right ventricle, and finally right-to-left shunt due to markedly increased right atrial and ventricular blood pressure.

S4 gallop is generally associated with increased ventricular hypertrophy and stiffness, still reported in patient with partial AVCDs (1,7). Because S4 gallop generally can be occurred with pressure overload during systolic period due to pulmonary hypertension and pulmonary stenosis (14), S4 gallop of this patient might be occurred as a result of increased right ventricular end-diastolic pressure due to a large left-to-right shunt, which was described in elsewhere (14).

The most common ECG findings of the partial AVCD are 'deep S waves' in leads I, II, III and aVF, and right axis deviation of the QRS due to left to right shunt (13). Bundle branch blocks on the ECG are also common, probably because the developmental defects interfere with normal growth of the cardiac conduction fiber (7,16). ECG findings of this case were distinctive of left anterior fascicular block (a positive QRS complex in lead I, negative QRS complex in aVF, and left axis deviation of the mean electrical axis) and atrial enlargement (tall P wave). Our interpretation for left anterior fascicular block in this case was there might be the disconnection of conducting fiber below left bundle branch due to developmental defects. A similar finding has been already reported in humans (12). Higher frequency of bundle branch block in the left anterior fascicle might be because the left anterior fascicle is thinner and more vulnerable than left posterior fascicle to disruption (12). The tall P wave in our case was also consistent ECG findings of AVCDs described in veterinary literatures (1,7).

The definitive diagnosis of the partial AVCDs is mostly made by the echocardiography. Definitive findings of the partial AVCDs are ostium primum defect on the right parasternal long axis view and the left apical four-chamber view, and a well developed cleft in the septal cusp of the mitral valve in the right parasternal short axis view (6,7,13). In addition, the possible abnormal findings are downward displacement of the atrioventricular valve, a cleft or dysplasia of the septal anterior tricuspid valve leaflets, and prolongation of the left ventricular outflow tract called 'goose neck' (1,7,10,11). Our echocardiogram clearly demonstrated ostium primum defect in inter-atrial septum and well developed cleft in the septal cusp of the mitral valve, which were most diagnostic findings in the echocardiography (7,13). In case that the direct evidence of cleft in the septal cusp of the mitral valve is not ready to identify, because of lower resolution echocardiography, the regurtitant flow from the mitral valve on the colour Doppler echocardiography may be a good alternative echocardiographic finding for the presence of cleft of the mitral valve (7,10,13), although it is not generally recommended. However, in our case, we clearly demonstrated the presence of cleft of the mitral valve, which was diagnostic finding for differentiating AVCDs from ASD.

Patients with partial AVCDs have the same pathophysiology as a large septal defect (7). The clinical signs of the partial AVCDs are mainly dependent on the shunt direction and the size of the defect of the ASD. Generally large defects (e.g. os primum ASD or AVCD) result in marked right ventricular eccentric hypertrophy, because the right ventricle is approximately 2 to 3 times as compliant as the left ventricle and thus the right ventricle can accommodate more blood volume than left ventricle if there is no interatrial septum (7). This often causes shunt-reversal and poor prognosis, if the right atrial and ventricular pressure surpasses the left ventricular pressure. However, in our case, early diuretic and ACEi therapy might significantly reduce right ventricular volume overload and markedly slow down the disease progression to shunt reversal. Recent study found the ratio of pulmonary blood flow to systemic blood flow (Q_p:Q_s) using pulsed-wave Doppler echocardiography had prognostic value for septal defects (8,13), since the Q_p:Q_s ratio reflecting pulmonic and aortic flow volumes is useful in evaluating the degree of the intracardiac shunts (e.g. ASD), although more studies are warranted for verification of this method.

Although recent development of cardiac intervention enables us to close small size septal defects, large septal defects seen in our case are generally incurable with interventional or surgical methods. However surgical trial to treat similar cases have been reported but showed bad outcome (11). Other surgical repairs of the partial AVCDs have also been reported in veterinary literatures (2,10). Because surgical methods need more technical development (10,11), our decision for managing this case was medical therapy focusing on the reduction of right ventricular volume overload. In veterinary literatures (1,7), the recommended medical treatments for similar cases, are ACEi, diuretics and vasodilators, which employed in this case although the owner eagerly wished more definitive treatment. Our group is currently considering treatment of this case using the combination of surgical repair with interventional method using septal occluder (e.g. amplatzer® device).

To the best of our knowledge, this is the first case report of partial atrioventricular canal defect of dog in Korea.

Acknowledgements

This study was supported by research grant from Korean Research Foundation (KRF-2007-331-E00369).

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