

Ebstein's Anomaly in a Beagle Dog

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ABSTRACT. An intact male beagle dog aged 1 year was referred because of shortness of breath, exercise intolerance and cardiac murmur. Based on the results from electrocardiography, thoracic radiography and echocardiography, the dog was diagnosed as Ebstein's anomaly. Although the orally administered digoxin, vasodilators and diuretics partially improved congestive signs, the dog became to be refractory and died 20 months after the diagnosis. Necropsy confirmed malformation and apical displacement of the basal attachment of tricuspid valve leaflets.

KEY WORDS: canine, Ebstein's anomaly.

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Echocardiography has become an essential examination for diagnosis of congenital heart disease (CHD) in small animals, allowing the diagnosis of uncommon or complex CHD. One of such diseases is malformation of the tricuspid valve, and which may be either tricuspid dysplasia or Ebstein's anomaly. The former is characterized by various abnormalities of the leaflets, chordae tendineae and papillary muscles, and the details have been described in veterinary literatures [11, 12]. On the other hand, Ebstein's anomaly is characterized by downward displacement of the basal attachments of the tricuspid valve leaflets [2]. Although 8 dogs with Ebstein's anomaly have been documented to our knowledge [4, 6, 9, 10, 13, 15], extremely brief descriptions lacking in clinical and/or pathological finding were included in these reports. Therefore, detail clinicopathological finding of this malformation is thought to be limited in dogs. The present paper describes the clinicopathological findings of Ebstein's anomaly in a dog.

An intact male beagle dog aged 1 year weighing 8.7 kg was referred to the Veterinary Medical Teaching Hospital, Nippon Veterinary and Animal Science University, because of shortness of breath and exercise intolerance. The referring veterinarian had suspected pulmonary stenosis based on cardiac systolic murmur and enlargement, and had treated with digoxin (0.01 mg/kg/day), furosemide (4 mg/kg/day), isosorbide dinitrate (40 mg/dog/day), and enalapril maleate (2.5 mg/dog/day). However, the congestive signs progressively became refractory.

On admission, the dog showed inactivity and normal appetite. The prevention of heartworm was complete. Physical examination revealed mild panting. Heart rate was 116 beats per minute, and a grade 4/6 regurgitant systolic murmur was auscultated over the tricuspid area. No arrhythmia was auscultated, and lung sounds were normal. Capillary refill time was 2 sec. The jugular veins were dis-

tended but not pulsated. Mild hepatomegaly was palpated, and the ascites were not detected. The results of routine blood work were within reference ranges.

An electrocardiogram (Fig. 1) revealed first-degree atrioventricular block with markedly prolonged P-Q segment

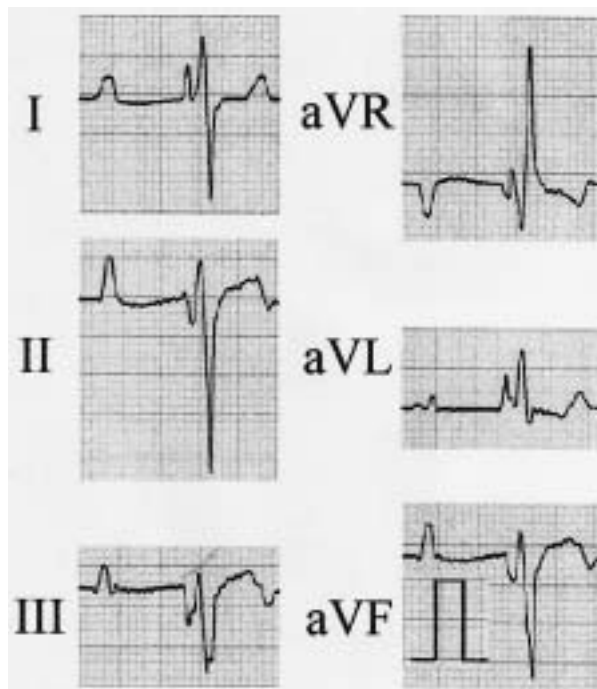


Fig. 1. Six lead electrocardiogram recorded from a dog with Ebstein's anomaly. Large and wide S waves in leads I, II, III and aVF, positive QRS deflections in leads aVR and aVL, and right axis deviation (100 degree) in addition to prolonged QRS duration (0.12 sec) are consistent with right branch bundle block. First-degree atrioventricular block is also noted (PQ interval: 0.22 sec).



Fig. 2. Two-dimensional echocardiography of a dog with Ebstein's anomaly. This image was the left parasternal apical four-chamber view. The right atrium (RA) markedly enlarged, and basal attachment of the tricuspid septal (SL) and posterior leaflets (PL) displace to the apex. RV: Right ventricle, LA: Left atrium. An arrowhead indicates the mitral valve.

(0.22 sec). Prolonged P wave (0.06 sec) and QRS complex (0.12 sec) durations were recorded. Large and wide S waves in leads I, II, III and aVF, positive QRS deflections in leads aVR and aVL, and right axis deviation (100 degree) were also noted, and these findings were consistent with right bundle branch block (RBBB) [7]. Thoracic radiography

(not shown) revealed marked enlargement of the right heart, thin pulmonary arteries and veins, and widened caudal vena cava. Lung field was normal.

Two-dimensional echocardiography revealed marked dilations of the right atrium and ventricle (Fig. 2). In the tricuspid valve, the basal attachments of the septal and posterior leaflets displaced apically, and the resultant tricuspid regurgitation was observed. Other cardiac malformation was not detected. Based on these results, the dog was diagnosed as Ebstein's anomaly.

Oral digoxin (0.01 mg/kg, bid), enalapril maleate (0.5 mg/kg, bid), isosorbide dinitrate (0.5 mg/kg, bid) and furosemide (0.5 mg/kg, bid) were administered, and the improvement of clinical signs was achieved 3 weeks after the diagnosis. However, intermittent vomiting and abdominal distention due to the ascites developed 3 months later, and low sodium diet was also instituted. Five month after the diagnosis, the dog showed severe exercise intolerance, cold extremities, prolonged capillary refill time (2–3 sec), weak and hypokinetic pulse, increased ascites, hepatomegaly and the distention of the peripheral veins. A grade 5/6 regurgitant systolic murmur was auscultated over the tricuspid area. Electrocardiographic finding was similar to that of the initial examination. Thoracic radiography revealed further enlarged cardiac silhouette. Echocardiography revealed hepatic congestion as well as marked dilation of the right atrium and ventricle. The dose of furosemide (1

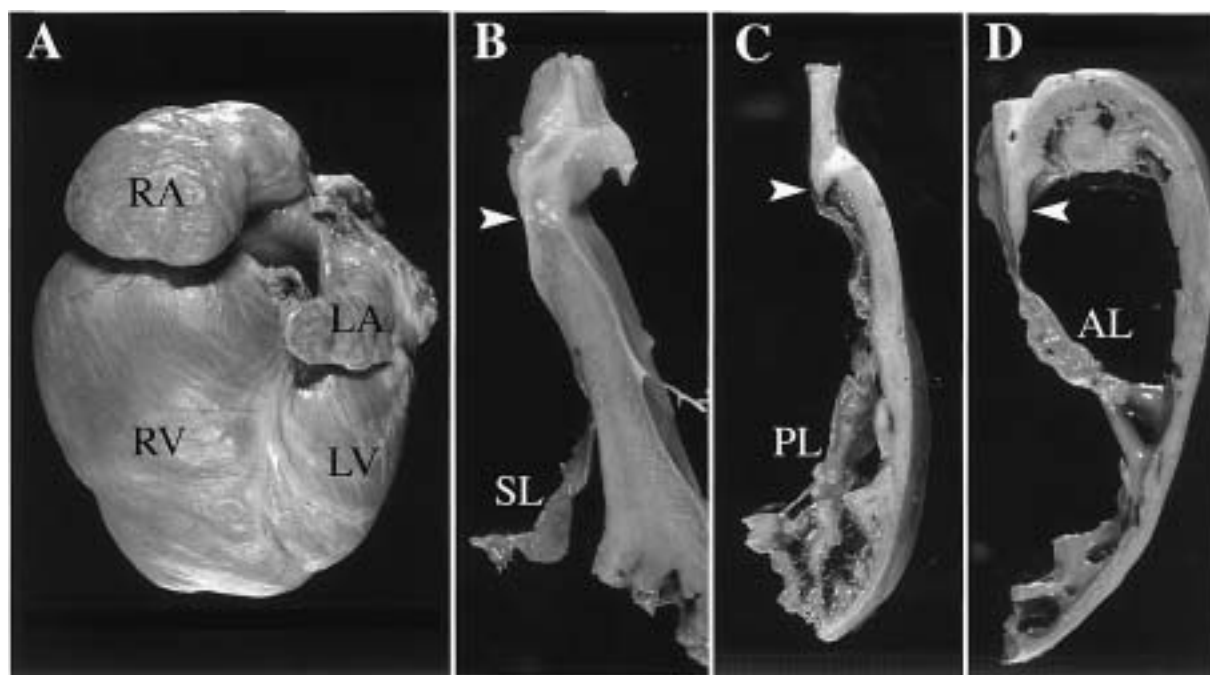


Fig. 3. Heart from a dog with Ebstein's anomaly, showing extreme enlargement of the right atrium and ventricle (A). Longitudinal sections of the ventricular septum (B), and the posterior and anterior walls of the right ventricle (C and D), showing misplaced and/or malformed tricuspid valve leaflets. The septal and posterior leaflets (SL and PL) are attached in part to the ventricular wall distal to the atrioventricular junction (an arrowhead), resulting in downward displacement of the functional tricuspid valve annulus into the right ventricle. The anterior leaflet (AL) inserts directly into the deformed papillary muscle. RA: right atrium, RV: right ventricle, LA: left atrium, LV: left ventricle. Scale=1 mm.

mg/kg, bid) was increased and spironolactone (0.5 mg/kg, bid) was added for the treatment. Although congestive signs were partially improved, the dog died 20 months after the diagnosis.

Pathological findings: Gross examination of the external surface of the heart revealed right-sided enlargement (Fig. 3A). On section, conspicuous dilatation of the right atrium and ventricle was present, and the tricuspid valve leaflets as well as the papillary muscles and chordae tendineae were totally abnormal. The valve leaflets were malformed and elongated. The upper half of the septal leaflet was plastered to the ventricular septum (Fig. 3B), the posterior leaflet was attached in part to the posterior ventricular wall (Fig. 3C), and the anterior leaflet arose from the atrioventricular junction but inserted directly into the deformed papillary muscles on the anterior ventricular wall without chordae tendineae (Fig. 3D). As a result of the downward displacement of the functional tricuspid valve annulus into the right ventricle, the inflow tract of the right ventricle lay between the atrioventricular junction and the origin of the displaced valve, which was apparently included in the right atrial cavity (i.e., the atrialized ventricle).

Ebstein's anomaly is uncommon in animals, and few canine cases had been reported [4, 6, 9, 10, 13, 15]. This malformation is characterized by the downward displacement of the attachment of the tricuspid valve, resultant tricuspid regurgitation and the presence of atrialized right ventricle [2]. In the present case, these findings were detected by echocardiography and confirmed by necropsy.

In human patients, RBBB is frequently associated with Ebstein's anomaly [8]. This arrhythmia was also observed in the present case. In general, RBBB might commonly result from structural heart disease, Chagas disease, heartworm disease, acute pulmonary thromboembolism and hypokalemia in dogs [7]. Although its mechanism could not be determined in the present case, Ebstein's anomaly should be included when making the differential diagnosis for canine RBBB.

The management of humans with Ebstein's anomaly requires medical and/or surgical treatment [3]. In severe heart failure, the creation of arterial shunt between systemic and pulmonary circulations, the Fontan procedure, and repair or replacement of the tricuspid valve may be considered. Although such procedures might also be a treatment of choice for the present dog, surgery was not selected because of the technical problems. Diuretics and digoxin are recommended for human with symptomatic Ebstein's anomaly. In addition to these drugs, vasodilators and low

sodium diet were also given to palliate cardiac overloading. As a result, congestive signs were partially improved, and no adverse reaction was noted in the present dog.

The etiology of Ebstein's anomaly has not been studied in veterinary medicine. In humans, maternal lithium intake has been suggested as a causal agent [14]. Dominantly inherited syndrome of Ebstein's anomaly and skeletal abnormalities has been documented [1]. In one case-control study, twins, family history of CHD, previous miscarriages, maternal exposure to benzodiazepine and varnishing were shown to be the risk factors of the malformation [5]. Histories related to maternal reproductive status, maternal chemical intake and familial occurrence could not be determined in the present case. Detailed investigation will be required to elucidate the etiology of Ebstein's anomaly.

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