

Ebstein's anomaly in a French bulldog

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Abstract

A one and a half years old male French bulldog weighing 9.50 kg was presented with the history of inappetence, lethargy, abdominal distension and exercise intolerance since last 2 days. The physical examination was done which revealed normal physiological parameters including temperature, mucus membrane color and capillary refill time except palpable precordial thrills, jugular distension on palpation, tachycardia and systolic murmurs on auscultation. Electrocardiography (ECG) was done which depicted ectopic foci with atrioventricular junctional tachycardia and right ventricular enlargement involving very small inverted P waves, deep S waves in leads I, II, III and augmented vector foot (aVF) and splintered QRS complexes. The dog was undergone chest radiography that revealed right atrial enlargement, increased sternal contact of heart on lateral view and a bulge at 9:00 o'clock to 11:00 o'clock depicted right atrial enlargement on dorso-ventral view. Lastly, echocardiography was done to arrive at a diagnosis confirming the Ebstein's anomaly as a form of tricuspid valve dysplasia including apical displacement of tricuspid valve leaflets, division of right ventricle into atrialized and functional portions, increased displacement index, increased apex-mitral annulus to apex-tricuspid annulus ratio, severe right atrial dilatation and tricuspid regurgitation. The dog was medically treated with diuretics, angiotensin converting enzyme inhibitors and inotropes and the owner was advised to put the dog on low sodium diet for 2 weeks. The dog has resolved clinical signs of right sided heart affection; but, suddenly collapsed at home. The owner denied for the necropsy of dog.

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Introduction

Tricuspid dysplasia is a congenital anomaly constituting about 2.00 - 7.00% of all the congenital cardiac malformations in dogs¹ including improper position or structure of valve leaflets, chordae tendineae or papillary muscles.² Ebstein's anomaly is a special case of tricuspid dysplasia whose prevalence is 2.90% according to one study³ in which tricuspid valve leaflets were apically displaced compared to the level of mitral annulus and there was a division of right ventricle (RV) into the functional and atrialized RVs leading to enlarged right atrium (RA) and tricuspid regurgitation ultimately leading to right sided heart failure signs.⁴ Labrador retriever is the most commonly affected breed.⁵

The present case report describes this anomaly in a young male French bulldog.

Case Description

A one and a half years old male French bulldog weighing 9.50 kg was presented to the Multi-Specialty Veterinary Hospital of Guru Angad Dev Veterinary and Animal Sciences University in Ludhiana, Punjab, north India, with the history of inappetence, lethargy, abdominal distension and exercise intolerance since last 2 days. There was no history of coughing and syncope episodes. The dog underwent thorough physical examination which revealed normal parameters including temperature, mucus membrane and capillary refill time except palpable precordial thrills, tachycardia and grade IV systolic murmurs on tricuspid area of heart auscultation. Systolic blood pressure was 90.00 mmHg which was measured by Doppler method (Vet-dop2, Model BF2, Vmed Technology, Mill Creek, USA). Chest radiography was done in lateral

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and dorso-ventral (DV) views. Lateral radiograph revealed globoid appearance of heart occupying 4.50 inter-costal space along with increased sternal contact of heart and mild pleural effusions (Fig. 1A). The DV view also revealed a bulge at 9:00 o'clock to 11:00 o'clock indicating right atrial enlargement (Fig. 1B).

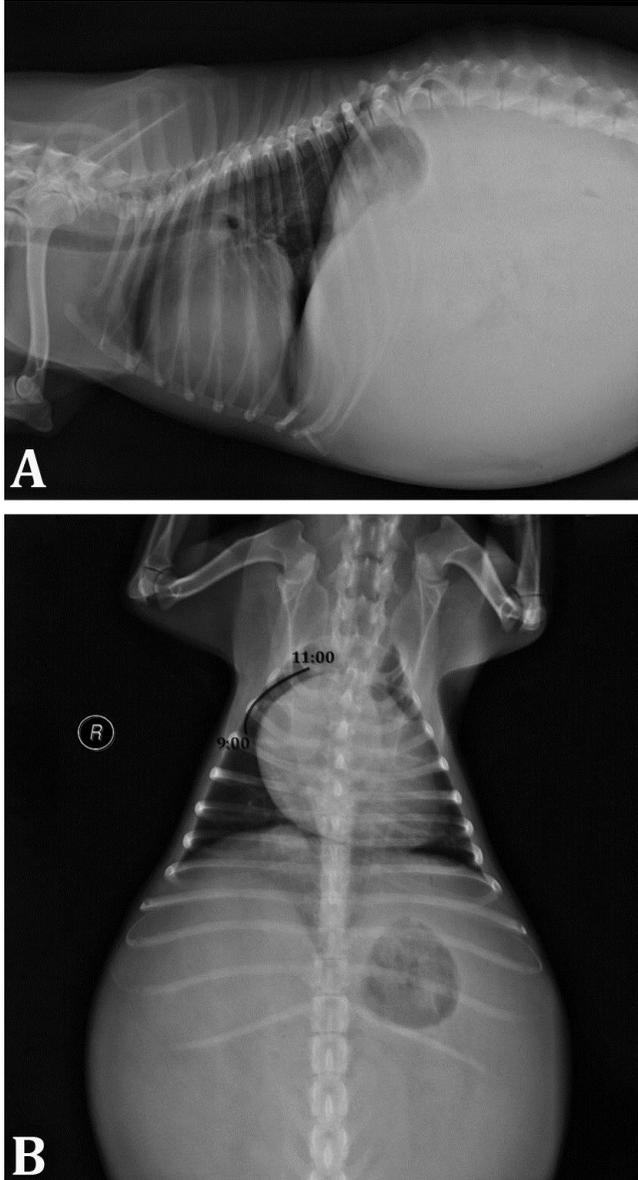


Fig. 1. A) Lateral chest radiograph depicting cardiomegaly (4.50 inter-costal space and 12 vertebral heart score) and increased sternal contact with mild pleural effusions. **B)** Dorso-ventral view of chest depicting a bulge at 9:00 to 11:00 o'clock (black curve line) confirming right atrial enlargement.

Electrocardiography (ECG) was done by putting the dog in right lateral recumbency using 6-channel electrocardiographic machine (Cardiart 8108; BPL Medical Technologies, Bangalore, India). The electrocardiogram revealed ectopic foci in atrioventricular (AV) junction with

AV junctional tachycardia which was indicated from very small inverted P wave (0.05 mV) just before forming R wave and right ventricular enlargement due to the presence of deep S waves in leads I (0.30 mV which was more than normal 0.05 mV), II (0.90 mV which was more than normal 0.35 mV), III (0.60 mV) and augmented vector foot (aVF; 0.80 mV), (Fig. 2). Splintered QRS complexes were also visible in lead III being indicative of right AV valve malformation (Fig. 2).

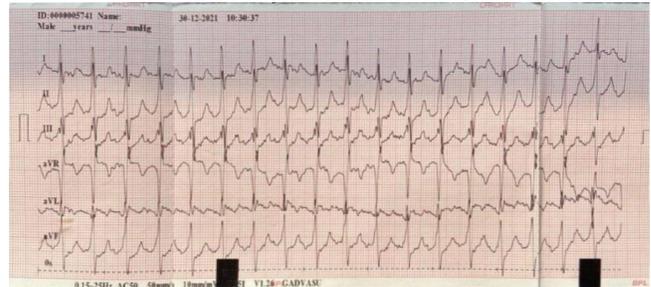


Fig. 2. Electrocardiography at 50.00 mm sec⁻¹ paper speed and 10.00 mm mV⁻¹ sensitivity showing very small inverted P waves, deep S waves in leads I, II, III and aVF suggesting junctional tachycardia with ectopic foci and right ventricular enlargement, respectively. Also, splintered QRS complexes in lead III suggest right atrioventricular valve malformation with tachycardia (heart rate: 250 bpm).

Hemato-biochemical picture revealed normal hemoglobin as well as total leukocyte and platelets counts except absolute neutrophilia with mild left shift. Increased gamma-glutamyl transferase (34.00 IU L⁻¹), total bilirubin (1.90 mg dL⁻¹) and albumin (4.80 g dL⁻¹) levels with normal creatinine values were noted. Ascitic fluid was of serosanguineous nature with total protein of 3.70 g dL⁻¹.

Then, final echocardiography was done through standard two-dimensional ECG mode, M mode and Doppler mode using GE Logiq P5 color Doppler machine (GE healthcare, Chicago, USA). Downward apical displacement of tricuspid annulus and morphology of valve leaflets along with chordae tendineae were evaluated in right parasternal long axis and left apical four chamber views. Right parasternal long axis view revealed significantly enlarged RA with small left atrium (LA) and left ventricle. When color Doppler was applied in the same view, severe tricuspid regurgitation was observed (Fig. 3A). Using this right parasternal view to assess the severity of Ebstein's anomaly, RA: LA was measured which was calculated to be 2.07 depicting that RA was transversely dilated because its ratio was more than one (Fig. 3B). Then, in left apical four chamber view, tricuspid valve leaflets were seen apically displaced dividing the RV into atrialized and functional portions of RV (Fig. 4). Displacement index was also calculated from the same view which is the distance between the hinge point of mitral valve leaflet to the hinge point of displaced tricuspid septal leaflet at end-diastole divided by body surface area of animal (Fig. 4).

Displacement index for this case was 30.40 mm m^{-2} which was very high. Also, ratio between the distance from apex to the septal anterior mitral leaflet hinge point (AM) to the distance from apex to septal displaced tricuspid leaflet hinge point (AT) was calculated which came out to be 1.26 that was greater than normal value of less than 1.00 (Fig. 4). Based on these indices, this dog came under grade 2 of Celermajer classification of Ebstein's anomaly severity.⁶ This is because there was marked displacement of valve leaflets, large parietal tricuspid leaflet which was in normal motion with atrialized RV and small functional portion of RV as clearly depicted from various indices that was measured.

After confirming the diagnosis, the dog underwent abdominocentesis and was given furosemide as diuretic (3.00 mg kg^{-1} ; PO, bid), pimobendan as positive inotrope (0.25 mg kg^{-1} ; PO, bid) and L-carnitine (50.00 mg kg^{-1} ; PO, bid) along with liver supplements for 2 weeks. Also, the dog was digitalized for 5 days on the digoxin ($5.00 \mu\text{g kg}^{-1}$)

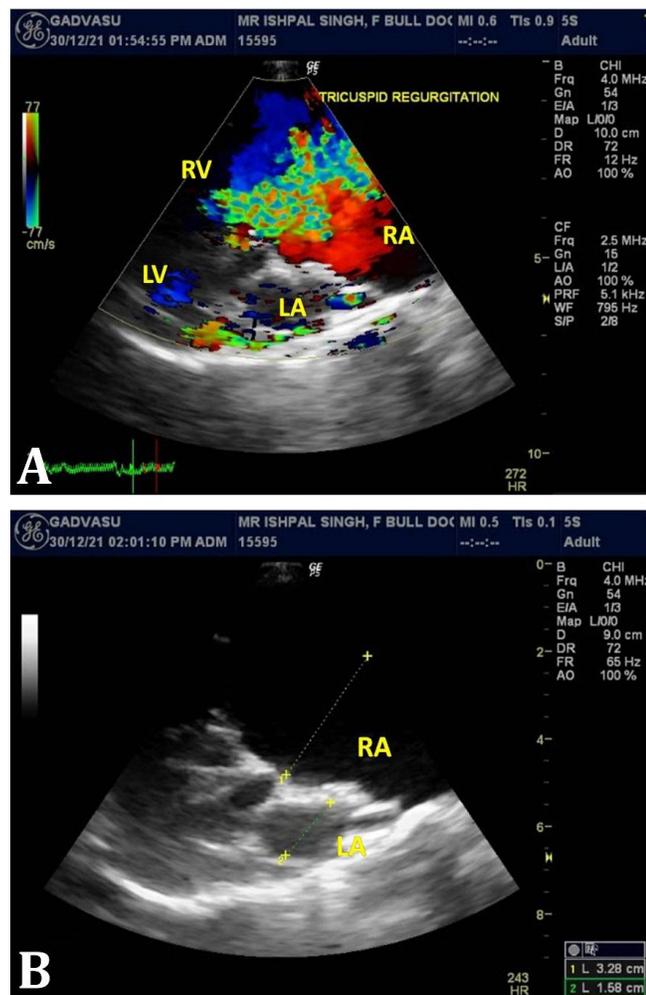


Fig. 3. A) Right parasternal long axis view depicting tricuspid regurgitation. LV: left ventricle, RV: right ventricle, LA: left atrium, and RA: right atrium. **B)** The ratio of RA: LA was 2.07 from right parasternal long axis view.

to relieve the congestive heart failure signs. The dog was improving with resolved ascites and normal activity within 2 weeks of treatment; but, unfortunately after 2 weeks, the dog collapsed suddenly and the owner denied for post-mortem examination.

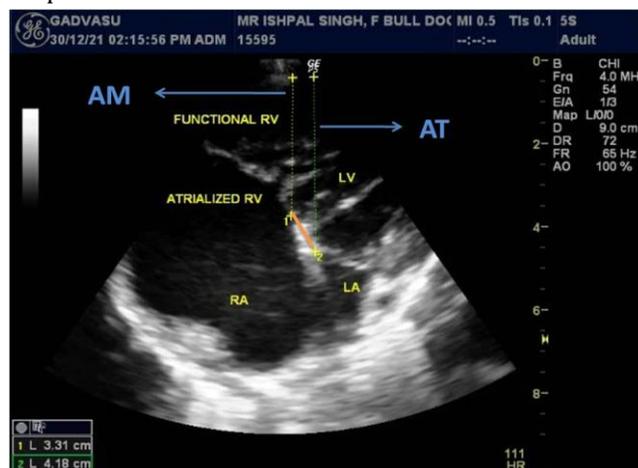


Fig. 4. Compartmentalization of right ventricle into functional and atrialized RVs due to tricuspid valve dysplasia being seen from left apical four chamber view. Displacement index was calculated by measuring the distance between hinge point of septal mitral leaflet and hinge point of septal displaced tricuspid leaflet and dividing the value by body surface area of dog (30.40 mm m^{-2}). Also, distance from apex to septal mitral annulus (AM) to distance from apex to displaced tricuspid leaflet annulus (AT) ratio was 1.26. LV: left ventricle, RV: right ventricle, LA: left atrium, and RA: right atrium.

Discussion

Tricuspid dysplasia accounts for approximately 7.00 to 7.50% of the congenital cardiac anomalies. While, Ebstein's anomaly as a typical case of tricuspid valve dysplasia is uncommon in dogs and cats.⁷ Though the most represented dogs are large breeds,³ small breeds like French bulldogs are also predisposed to this anomaly.⁸ The etiology of this anomaly is not clearly understood with dominant inheritance along with reduced penetrance and chromosome 9 susceptibility locus particularly in Labrador retriever dogs.⁵

The presented case reported a one and a half years old French bulldog with the signs of right sided congestive heart failure with palpable precordial thrills and tricuspid systolic murmurs that were revealed in some studies.³ Chest radiograph revealed severe right atrial enlargement as indicated by rounding of cardiac silhouette and 9:00 o'clock to 11:00 o'clock bulge on DV or ventro-dorsal views.⁹ Also, in severe tricuspid valve dysplasia, globoid appearance of heart is also visible along with pleural or pericardial effusions.¹⁰ Electrocardiographic findings are variable in cases of Ebstein's anomaly varying from typical presence of tall P waves to right bundle branch block, splintered QRS complexes and

various types of arrhythmias including atrial tachycardia and AV junctional tachycardia.^{4,11} Electrocardiogram in the presented case depicted ectopic foci with AV junctional tachycardia due to very small inverted P waves¹¹ and right ventricular enlargement being indicated by deep S waves in leads I, II, III and aVF. Splintered QRS complexes as detected in this case, suggest the conduction disturbances being usually present in right AV valve malformation and tricuspid dysplasia.¹² As depicted in various studies, ECG is not very sensitive in the heart chambers enlargement detection. Therefore, absence of peaked P waves as observed in this case cannot rule out this anomaly.¹⁰

Therefore, the echocardiography as a confirmatory technique for many congenital cardiac anomalies is warranted. Typical right atrial dilatation, atrialization of RV due to downward displacement of tricuspid valves and tricuspid regurgitation were found as depicted in other studies.^{3,13} Also, various echocardiographic indices being used in humans were done in the present study including displacement index, RA : LA, and AM : AT as measured by Chetboul *et al.*¹³ being indicators of Ebstein's anomaly as a special case of tricuspid valve dysplasia. Normal RA : LA is 0.60 - 1.02 and normal displacement index and AM : AT should be < 8.00 mm m⁻² and < 1, respectively.¹³ In the present study, RA : LA was 2.07, displacement index was 30.40 mm m⁻² and AM to AT ratio was 1.26, confirming the grade two of Ebstein's anomaly.

One should differentially diagnose the Ebstein's anomaly, whenever right sided heart failure signs with or without murmurs are evident with cardiomegaly on radiographs and normal or abnormal ECG. The most important differential diagnosis includes tricuspid dysplasia in which there is a normal insertion of tricuspid leaflets at AV ring and AV junction as a origin of tricuspid regurgitant jet.¹⁴ While, in Ebstein's anomaly, there is a presence of abnormally displaced tricuspid valve leaflet with the origin of regurgitant jet below the AV valve.¹⁴ The other less common differential diagnoses include tricuspid valve endocarditis, prolapse of tricuspid valve, right ventricular cardiomyopathy and pulmonic valve insufficiency - pulmonary hypertension. All these differentials in the present study were ruled out due to the presence of marked displacement of tricuspid septal leaflet (> 8.00 mm m⁻²) as well as elongated and redundant anterior tricuspid valve leaflets.

Treatment for Ebstein's anomaly in humans is mainly surgical intervention along with the medical management of right sided heart failure signs.¹⁵ However, in veterinary medicine, surgical intervention is not prevalent due to technical deficits. Prognosis of the patients diagnosed with this anomaly is variable.¹⁶ In the present study, poor prognosis of the dog was due to the presence of morphological changes of valves along with the conduction disturbance because of junctional tachycardia

in spite of the medical treatment. Therefore, this is an unusual congenital Ebstein's anomaly as a typical form of tricuspid valve dysplasia in a young male French bulldog with complete radiographic findings, ECG interpretation and echocardiography.

Acknowledgments

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Conflict of interest

The authors declare no conflict of interest.

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