Common Atrium or Atrioventricular Septal Defect: What Is in a Name?

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INTRODUCTION

Atrioventricular septal defect (AVSD) is a well-described group of congenital heart conditions. The hallmark features for all types of AVSD are absence of atrioventricular (AV) septal structures and a common AV junction guarded by a common AV valve possessing five leaflets.[1] The most frequently observed form of an AVSD with two valve orifices is an isolated primum defect between the lower edge of the interatrial septum (IAS) and the AV valve leaflets (partial AVSD).[1] The term "common atrium" has been used to denote the condition characterized by complete or near-complete absence of the IAS, with or without AVSD.[2,3] It is an uncommon disorder in humans that has been reported as individual case reports or as small case series. Common atrium and AVSDs are among the uncommonly reported congenital heart malformations in the domestic dog, but the rarity of reports of common atrium in dogs may simply be reflective of the inconsistent terminology used to describe this lesion. In humans, cases of common atrium are typically syndromic and have been reported in patients with heterotaxy, Ellis-van Creveld syndrome, and complete AVSD with and without concurrent Down syndrome.[4] Due to frequent occurrence of concurrent AV valve malformation, common atrium has been suggested to be a form of partial AVSD. However, the available literature in human patients suggests that it can occur as an isolated entity, without concurrent AVSD.[2] Here, we report a case of a young dog that presented for subclinical (asymptomatic) murmur evaluation, in which features of common atrium and partial AVSD were both observed using echocardiography. This case highlights that the constellation of congenital malformations that deviate from the usual morphology may pose challenges in assigning a single unambiguous term.

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CASE PRESENTATION

A 1.5-year-old, intact, female Belgian Tervuren dog weighing 17.6 kg was presented to the Atlantic Veterinary College Teaching Hospital for evaluation of a heart murmur prior to undergoing general anesthesia for elective ovariohysterectomy. The dog did not have any history of weakness, exercise intolerance, collapse, or respiratory signs. However, the dog was reported to be underweight by the primary care veterinarian approximately 3 months before presentation. The dog had been receiving clomipramine hydrochloride for presumed generalized anxiety disorder. Physical examination revealed a grade 5/6 left basilar systolic and a grade 1-2/6 left basilar diastolic murmur and a split second heart sound. The dog was moderately underweight with moderate muscle loss. The rest of the physical examination was unremarkable. Results of a complete blood count and serum chemistry were within reference ranges.

A transthoracic two-dimensional (2D) and Doppler echocardiographic study (Video 1) was performed with the patient unsedated. The IAS was not evident on several of the 2D imaging planes, suggesting a single atrial chamber (Figure 1). Upon careful interrogation, a small strand of tissue was noted in the dorsal wall of the common atrial chamber in some views, possibly representing a remnant of the IAS (Figure 1E). There was malformation of the AV valves (Video 1). Specifically, there was a common AV valve with septal leaflet (bridging leaflet) inserting at the same level on the crest of the interventricular septum (Figure 1A-B) and with the left-side portion of the AV valve being trifoliate in appearance (best visualized in the right parasternal short-axis left ventricle [LV]–right ventricle [RV] view; Video 1 and Figure 2A). These findings were consistent with the presence of a common AV annulus with two separate orifices (left and right AV orifices). The RV (end-diastolic diameter of 34 mm; Figure 1A–B, 2B-C) and main and branch pulmonary arteries were markedly dilated, suggesting marked right-sided volume overload (Video 1). The ratio of pulmonary (Qp) to systemic (Qs) flows was indirectly estimated by combining 2D echocardiography and pulsed-wave Doppler modes.[5] The Qp/Qs ratio was 5.1:1, indicating marked pulmonary overcirculation due to left-to-right shunting (Table 1). The left ventricular (LV) end-diastolic (33 mm) and end-systolic (24 mm) dimensions, LV wall thickness, and systolic function (fractional shortening 27%) were normal when compared with published allometric weight-based normal reference ranges.[6]

Color Doppler imaging showed moderate left and right AV valve regurgitation (Video 1). The direction of shunting in the atrial chamber could not be accurately determined because of the turbulent jets of left and right AV valve regurgitation and the atrial chamber inflow. Interventricular communication was not noted. The aortic and pulmonic valves appeared normal. On the right parasternal four-chamber imaging planes tipped to accentuate the apical portion of the ventricles, a trivial amount of pericardial effusion was noted (Figure 2D), which was considered to be within the physiological amount. On

VIDEO HIGHLIGHTS

Video 1: Transthoracic two-dimensional echocardiogram from the right and left parasternal windows with and without color Doppler showing the presence of a large single atrial chamber, malformations of the atrioventricular valve (a common atrioventricular annulus with two separate orifices), marked right ventricular dilatation, and moderate left and right atrioventricular valve regurgitation.

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continuous-wave spectral Doppler interrogation, the right AV valve regurgitation (Vmax = 3.6 m/sec) and pulmonic regurgitation (Vmax = 2.3 m/sec) jet velocities (in combination with the 2D features) suggested high probability for the presence of at least a moderate degree of pulmonary hypertension (PH).[7] The left AV valve inflow pattern was normal. Mild pulmonic regurgitation was present.

Two terms were considered to describe the echocardiographic findings in this dog: (1) partial AVSD with a large ostium primum atrial septal defect (ASD) and possibly a concurrent ostium secundum ASD and (2) common atrium with concurrent AVSD. These malformations resulted in secondary marked right-sided cardiac enlargement, pulmonary overcirculation, and PH.

DISCUSSION

The present case report describes echocardiographic findings in a young dog, which comprised near-complete absence of the IAS or, at most, its representation by a small strand of tissue present in the cephalad wall of the atrial chamber, along with absence of interventricular communication and malformation of the AV valves (common AV valve with two orifices and a trifoliate left AV valve). This case illustrates the indispensable role that echocardiography plays in the diagnosis of congenital heart diseases (CHDs), highlights the inconsistencies in terminology and nomenclature of certain congenital heart malformations in the literature, and emphasizes that the constellation of congenital malformations that deviate from the usual morphology may pose challenges in assigning a single unambiguous diagnosis.

In the literature, there is considerable nosologic confusion regarding the use of the terms "common atrium," "single atrium," and "AVSD." The term "AVSD" is a generic name for a group of anomalies characterized by absence of AV septal structures and consequently possessing a common AV junction guarded by a common AV valve.[8] This common valve can have a single (common) orifice or can be divided into two orifices by a tongue of valve tissue joining the bridging leaflets.[1] The spectrum of AVSD can range from partial (shunting confined at either the atrial or the ventricular level with one common annulus and two separate AV orifices), intermediate (shunting at both atrial and ventricular levels with one common annulus and two separate orifices), or complete (shunting at both atrial and ventricular levels with one common AV annulus and one single AV orifice) AVSD.[1,3,9] An AVSD can occasionally exist in the absence of any ASD or interventricular communication.[1] Even this nomenclature and subdivision of the varying morphology of AVSDs, which is widely used, is felt by some researchers to lack specificity.

Based on Rastelli *et al*'s[10] observations in 15 human patients and previous other reports stating that the complete absence of atrial septum always occurs with concurrent AV valve abnormalities, the common atrium was initially considered a form of AVSD. However, Levy *et al*,[3] in 1974, followed by several other investigators, reported cases of complete absence of the atrial septum without malformation of the AV valves, indicating that this condition may exist alone as a specific entity without an AVSD. They recommended that the term "single atrium" be used to denote the condition characterized by (1) complete absence of the atrial septum, (2) absence of malformation of the AV valves, and (3) absence of interventricular communication. They suggested that the term "common atrium" should be used to denote the condition of complete absence of the atrial septum, accompanied by malformation of the AV valves, with or without interventricular communication. To standardize the nomenclature for CHD across the globe, the International Nomenclature Society linked its efforts with those of the World Health Organization to obtain a globally accepted nomenclature tree for CHD with the 11th iteration of the International Classification of Diseases. According to this publication,[3] common atrium with separate AV junctions has been defined as "a congenital cardiovascular malformation in which there is complete or near-complete absence of the interatrial septum" and single atrium and atrium communis are denoted as synonyms. Common atrium with common AV junction has been defined as "a congenital cardiac malformation in which there is complete absence of the interatrial septum in the setting of a common atrioventricular junction (common atrioventricular canal)."[3] The echocardiographic findings in the dog in this report have features that fit partially into both of these categories, that is, near-complete absence of the IAS but with a common AV junction.

Atrioventricular septal defect is a rare CHD in domestic animals, with a reportedly higher occurrence in cats than in dogs.[11,12] Common atrium is an even more rarely reported CHD in veterinary medicine. However, this may simply be reflective of the inconsistent terminology used to describe this lesion and may not be a representation of its low prevalence both in the canine population and in the literature. Considering various definitions that have been used or proposed in the human and veterinary literature to describe AVSD or common atrium, there are at least two possible terms that could be applied to the echocardiographic findings observed in the dog of the present case report: (1) partial AVSD with a large ostium primum ASD and possibly a concurrent ostium secundum ASD (to account for the lack of IAS tissue) and (2) common atrium with AVSD. Potentially in favor of the second term are multiple human studies suggesting that, despite the AV septal deficiency and the presence of unequivocal shunting at the atrial level, the atrial septum is usually a well-defined structure in the setting of AVSD.[13] Similarly, in the largest case series pertaining to AVSD in the veterinary field, many cats with AVSD appeared to have an atrial communication yet had normal to near-normal atrial septal lengths.[12] These findings are not surprising because embryologically, the shunting at the atrial level in partial AVSD has been described to be due to the deficiency of the muscular portion of the AV septum (present between the offset hinges of the AV valve leaflets and the fibrous AV septum) and not due to the deficiency of the IAS itself.[13] The dog in the present report has near-complete deficiency of the IAS. If the echocardiographic findings in this dog are considered a form of partial AVSD, it would have to be

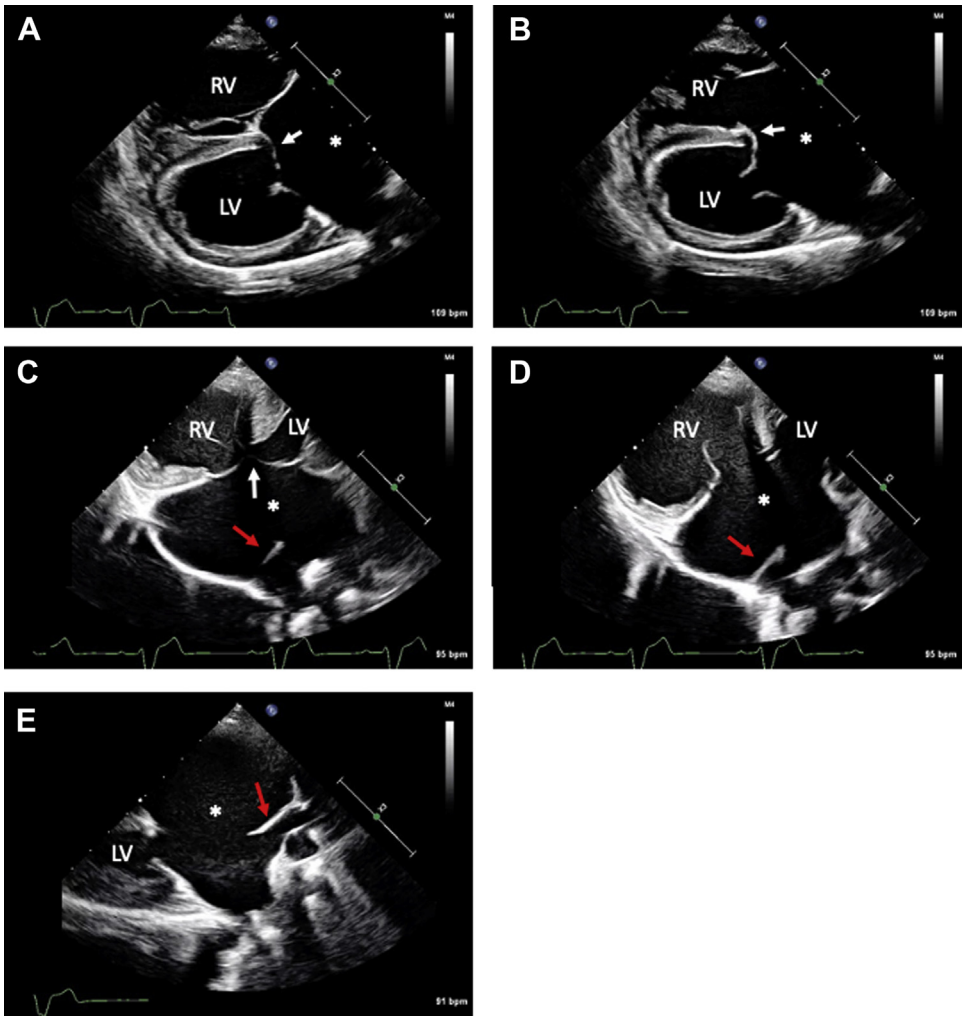

Figure 1 Echocardiographic images from a young dog showing a large atrial chamber (*asterisk*) with near-complete absence of the IAS. The IAS is represented by a thin strand of tissue (*red arrows*) attached as a remnant to the roof of the common atrial chamber. The left and right AV valve leaflets are at the same level (*white arrows*). The RV is markedly dilated. The LV dimensions are within the normal range. **(A)** Right parasternal long-axis four-chamber imaging plane in systole with AV valve closed. **(B)** The same image as in panel A recorded in diastole with AV valve open. **(C)** Left parasternal apical four-chamber imaging plane in systole. **(D)** The same image as in panel C recorded in diastole. **(E)** Modified right parasternal four-chamber view projecting the common atrial chamber.

Table 1 Results of Doppler echocardiography for Qp/Qs estimation

	Main pulmonary artery	Aorta
Diameter measured at end systole, cm	2.0	1.5
Systolic flow velocity-time integral, cm	36.2	12.5
Flow volume, mL	113.7	22.1

Measurements reflect the mean of three cardiac cycles.

with a markedly large ostium primum ASD, concurrent ostium secundum ASD, or both. In this scenario, this case would represent a form of partial AVSD in which the IAS is not a well-defined structure, in opposition to that described above.[12,13] This case could be considered a form of partial AVSD with features that deviate from the usual morphology.

Correct and consistent nomenclature for CHD is necessary for precise and accurate communication between colleagues and is crucial for accurate surgical planning and to predict the outcomes of surgical repair. Names are also essential in research, including retrospective studies, and teaching. It appears from the existing controversies surrounding the nomenclature of several CHDs that in patients with congenital heart malformation, we may not be able to rely entirely on individual names for particular conditions because of the presence of unusual features or associated anomalies. In these cases, a complete and accurate description in the diagnosis may be more accurate than attempting to fit them under a general term. The dog in this report can be described as having echocardiographic features of AVSD with a large interatrial communication. Whether the interatrial communication in this dog represents a form of common atrium or a large ostium primum defect and concurrent ostium secundum defect is debatable. Regardless of the terminology given to the findings in this dog, these congenital malformations resulted in secondary marked right-sided cardiac enlargement, pulmonary overcirculation, and PH.

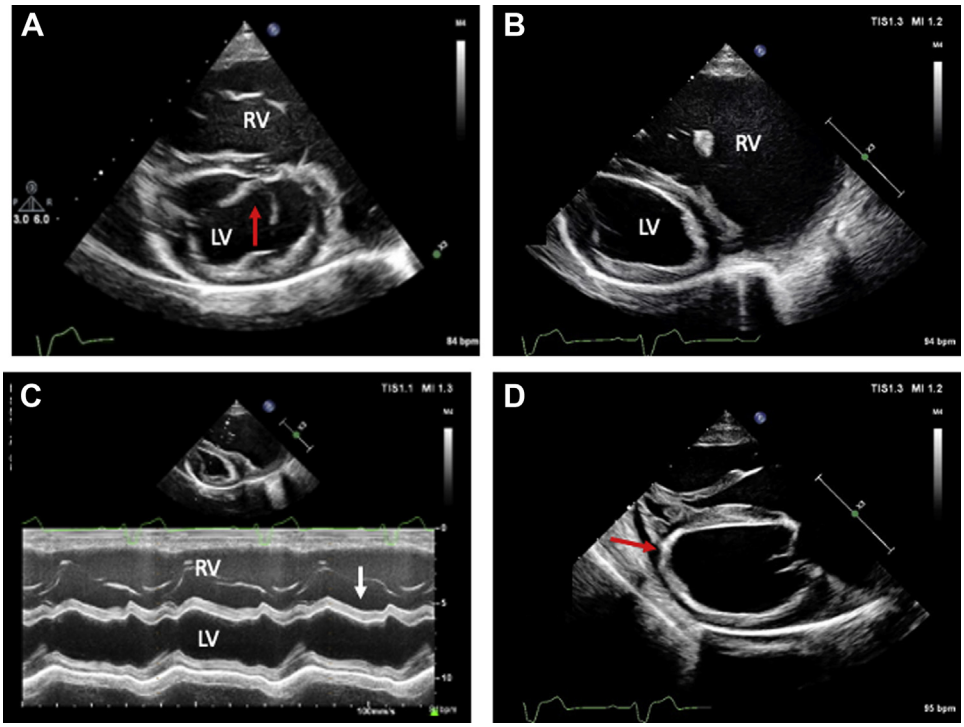

Figure 2 Echocardiographic images illustrating abnormalities detected in a young dog with a large interatrial communication and common AV junction. **(A)** Right parasternal short-axis (RPS SAx) transventricular (LV-RV) view shows a common AV valve with a trifoliate left AV valve (*red arrow*). **(B)** RPS SAx LV-RV view showing marked right ventricular dilation. **(C)** M-mode obtained from RPS SAx LV-RV view showing marked right ventricular dilation and diastolic flattening of the interventricular septum (*white arrow*). **(D)** Scant amount of pericardial effusion (*red arrow*) is noted on the RPS long-axis imaging plane.

The hemodynamic consequences in patients with common atrium or AVSD with atrial level communication are reported to be similar to those with large ASDs.[10] The amount of shunting that occurs across large ASDs depends primarily on the compliance of the two ventricles in diastole. Because the RV is approximately two to three times as compliant as the LV, in the absence of other cardiac abnormalities, large ASDs result in left-to-right shunting.[14] In this report, the combination of AV valve regurgitation and atrial inflow within the atrial chamber made it difficult to detect the direction of shunting. However, marked right-sided volume overload, lack of evidence supporting right-to-left shunting (e.g., lack of erythrocytosis, cyanosis), and Doppler-derived Qp/Qs ratio (5.1:1) all indicated left-to-right shunting. An agitated saline contrast study to further evaluate the presence of right-to-left shunting was not performed in this case.

Humans with common atrium frequently have concomitant abnormalities of systemic and pulmonary venous drainage.[15] In the previous case report of common atrium in a dog, persistent left cranial (superior) vena cava and anomalous systemic venous return were noted.[1] In the present report, anomalous venous drainage was not suspected based on the echocardiographic evaluation.

Treatment for children and young adults with common atrium or AVSD consists of surgical correction under cardiopulmonary bypass, which is recommended early in life due to risk for early development of pulmonary venous obstructive disease.[4] Surgical correction was not selected in this dog primarily because of the lack of widespread veterinary experience with such procedures. Due to the lack of clinical signs, no cardiac medications were prescribed. Follow-up echocardiographic examination after 5-6 months was recommended to assess remodeling and hemodynamics. There were no further updates from the owners at the time this report was written.

CONCLUSION

Common atrium and AVSD are among rare CHDs in domestic animals. A large interatrial communication with concurrent AV valve malformation may occur in dogs with a loud murmur and without overt clinical signs. The diagnosis can be made on the basis of 2D and Doppler echocardiographic imaging. The present case report describes a constellation of echocardiographic findings of near-complete absence of the IAS, a common AV junction, and absence of interventricular communication in a young dog. These malformations resulted in secondary marked right-sided cardiac enlargement, pulmonary overcirculation, and PH. These findings could be termed partial AVSD with a large interatrial communication or common atrium with AVSD. This case report is one example where a globally accepted nomenclature system for veterinary patients with complex CHDs could be a valuable resource to minimize inconsistencies in terminology.

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SUPPLEMENTARY DATA

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