Anomalous Left-to-Right Shunting Vessel between the Ascending Aorta and Right Pulmonary Artery and Concurrent Left Peripheral Pulmonary Artery Stenosis in a Dog

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INTRODUCTION

In dogs, the most common congenital heart disease characterized by a continuous murmur is a patent ductus arteriosus (PDA). Less common causes of a continuous murmur include aorticopulmonary windows, ruptured aneurysms of the sinuses of Valsalva, coronary artery fistulae, systemic-to-pulmonary arteriovenous fistulae, bronchial collateral vessels, and pulmonary vessels arising from a truncus arteriosus, or a continuous murmur may reflect turbulent blood flow in major arteries (such as in an aortic coarctation or severe branch pulmonary artery stenosis). We report an unusual case of an anomalous vessel between the ascending aorta and right pulmonary artery and a left peripheral pulmonary artery stenosis and speculate on possible embryologic origins of the former.

CASE PRESENTATION

A 4-month-old, male, intact Bulldog was referred to the University of California, Davis, Veterinary Medical Teaching Hospital for evaluation of an incidental heart murmur; the dog had no clinical signs at home. A grade V/VI left basilar continuous murmur and bounding femoral pulses were noted; the remainder of the physical examination was within normal limits for the breed. Three-view thoracic radiography showed generalized cardiomegaly with specific left atrial enlargement, an enlarged left caudal lobar pulmonary artery, and a mild bronchointerstitial and vascular pattern consistent with pulmonary overcirculation (Figure 1).

Transthoracic echocardiography (EPIQ 7C; Philips Medical Systems, Andover, MA) revealed severe left ventricular eccentric hypertrophy (normalized left ventricular internal diameter in diastole 2.10 cm/kg$^{0.299}$; reference range, 1.19–1.63 cm/kg$^{0.299}$), severe left atrial enlargement (normalized left atrial diameter measured in long-axis from the right parasternal four-chamber view at end-systole 1.83 cm/kg$^{0.309}$; reference range, 1.19–1.57 cm/kg$^{0.309}$), relative systolic dysfunction (fractional shortening 35%; normalized left ventricular internal diameter in systole 1.11 cm/kg$^{0.387}$; reference range, 0.50–0.92 cm/kg$^{0.387}$), and normal mitral valve leaflets with mild central regurgitation likely secondary to annular deformation (Figure 2A). Turbulent, continuous left-to-right flow near the junction between the main and branch pulmonary arteries was identified using color Doppler, with a peak velocity of 4.4 m/sec noted on continuous-wave Doppler (Figures 2B-2D, Videos 1 and 2). Although similar to the appearance of a PDA, the shunting vessel joined the pulmonary artery at an angle atypical for a PDA. No other shunting or valvular lesions were identified.

Transesophageal echocardiography and single-plane fluoroscopy-guided selective angiography were performed under anesthesia in preparation for intravascular occlusion of the shunting vessel. Color Doppler on transesophageal echocardiography identified the vessel with continuous left-to-right shunting (with a peak velocity of 4.1 m/sec on the basis of continuous-wave Doppler). However, the orientation was very peculiar in that the shunting vessel approached its pulmonary artery connection via the ventral aspect as opposed to the dorsal aspect, as expected for a PDA (Figure 3, Video 3). The origin of the vessel was not readily appreciated. A 6 Fr × 55 cm Ansel Top Cook Check-Flo introducing sheath (Cook Medical, Bloomington, IN) was placed in the right femoral artery. Initial injection of 10 mL of iodinated contrast (Omnipaque 300; GE Healthcare, Little Chalfont, United Kingdom) and 2 mL of saline in the proximal descending aorta failed to document a PDA, although partial opacification of an anomalous vessel was noted at the level of the aortic root and ascending aorta. The catheter was advanced, and an aortic root injection was subsequently performed by hand; although a Check-Flo introducing sheath was used for this purpose, substituting an angiographic catheter would have been ideal to optimize study quality and minimize possible subintimal damage. Nevertheless, the study confirmed an anomalous vessel arising from the ascending aorta connecting to the pulmonary artery (Figure 4, Video 4). A severely dilated left pulmonary artery was also identified. Two distinct coronary artery origins (left and right) were identified and were normal in appearance. A 0.035-inch HiWire (Cook Medical) was then advanced through the Check-Flo introducing sheath into the aortic root and was successfully placed across the anomalous vessel into the right pulmonary artery. However, when attempting to advance the Check-Flo catheter, the HiWire whipped back into the aortic root and would not maintain position across the anomalous vessel. This prevented the Check-Flo catheter from being maneuvered across the anomalous vessel. The intravascular occlusion procedure was then aborted.

To further evaluate the anatomy, computed tomographic images were acquired with a helical 16-slice scanner (LightSpeed, 16 helical

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VIDEO HIGHLIGHTS

Video 1: Transthoracic two-dimensional echocardiographic video of the right parasternal short-axis basilar view with color Doppler comparison of the continuously shunting anomalous vessel near the junction between the main and branch pulmonary arteries.

Video 2: Transthoracic two-dimensional echocardiographic video of the left parasternal short-axis basilar view with color Doppler comparison of the continuously shunting anomalous vessel inserting near the origin of the right pulmonary artery.

Video 3: Transesophageal two-dimensional echocardiographic video with color Doppler comparison of the continuously shunting anomalous vessel inserting into the ventral aspect of the pulmonary artery.

Video 4: Aortic root angiography with visualization of the anomalous vessel and an enlarged left pulmonary artery.

View the video content online at www.cvcasejournal.com.

scanner; GE Medical Systems, Milwaukee, WI). Iodinated contrast medium (Isovue-370; Bracco, Milan, Italy) was administered intravenously with a dose of 880 mg I/kg, and multiple contrast-enhanced series were sequentially obtained immediately after contrast administration. A three-dimensional volume rendering was also created. A tortuous, anomalous vessel was identified originating from the dorsal aspect of the ascending aorta and coursing slightly craniodorsally and toward the left (Figures 5 and 6). It inserted into the right pulmonary artery from the right cranial aspect at a level just distal to the main pulmonary artery. The diameter of the anomalous vessel tapered as it coursed toward the right pulmonary artery, where it terminated into a focal narrowing with a small ostium (diameter 0.5 cm). Additionally, there was a thin band of tissue encircling the left pulmonary artery near its origin from the main pulmonary artery causing focal attenuation of the luminal diameter (Figures 5 and 6). Distal to this, there was moderate to marked asymmetric dilation of the left pulmonary artery. This was interpreted as left peripheral pulmonary artery stenosis with poststenotic dilation.

Surgical ligation of the anomalous vessel was discussed with the owners, but the procedure was declined. The dog was prescribed oral benazepril 0.5 mg/kg twice daily and oral pimobendan 0.25 mg/kg twice daily. At the time of writing, the dog remains asymptomatic.

DISCUSSION

This case report highlights an especially rare congenital lesion resulting in a continuous murmur in a dog. Despite their prevalence in dogs as a species, PDAs are not noted to be a common finding in the Bulldog breed, and echocardiography provided further support for the lesion's atypical nature. Although similar cases have been reported in dogs, the anomalous vessel has an unclear embryonic basis.[1,2] Its origin at the ascending aorta distinguishes it from PDAs and systemic-to-pulmonary arteriovenous fistulae (also known as systemic-to-pulmonary collaterals), both of which typically originate at the descending aorta.[3] Among the possible communication abnormalities between the

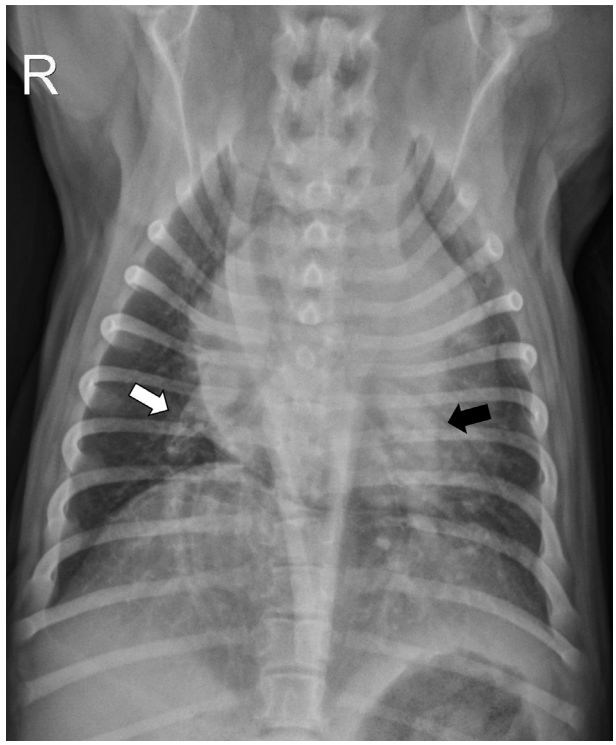

Figure 1 Thoracic radiograph, dorsoventral projection. The right caudal lobar pulmonary artery (*white arrow*) and the left caudal lobar pulmonary artery (*black arrow*) are highlighted, demonstrating notable enlargement of the latter.

ascending aorta and the pulmonary artery, most of the limited case reports in dogs are of aortopulmonary windows.[4,5] Aortopulmonary windows are characterized by an opening without significant length or tortuosity to the communicating channel.[6] Another consideration is coronary artery fistulae, which can occur between the coronary arterial system and the cardiac chambers (coronary cameral fistulae), the pulmonary arteries, or the venae cavae. Coronary-to-pulmonary artery fistulae have been reported in dogs and often manifest as dilated, tortuous vessels connecting the ascending aorta to the pulmonary artery. In this case the main coronary arteries were readily documented as normal on angiography.[7] An anomalous origin of the pulmonary artery is implausible given that the left and right pulmonary arteries are seen distinctly arising from the main pulmonary artery.

The most plausible diagnosis may be the presence of a persistent fifth aortic arch (PFAA), a rare congenital anomaly that has not been officially recognized in dogs or cats. During embryonic development in mammals, a total of six pharyngeal arches are formed and modified, with the fifth arch normally being evident only for a short duration. There are three types of PFAA: double-lumen aortic arch with both lamina patent (type I), atresia and interruption of the superior arch with patent inferior (persistent fifth) arch (type II), and systemic-to-pulmonary arterial connection arising proximal to the first brachiocephalic artery (type III).[8,9] Type I[8] and type II[9] PFAAs have been reported to be the more common forms in humans. In our case, type III PFAA is a plausible differential diagnosis for the anomalous vessel, although historically the existence of a fifth aortic arch is controversial, prompting a stricter proposed definition for a PFAA, in which the channel "must be confined to the extrapericardial space,

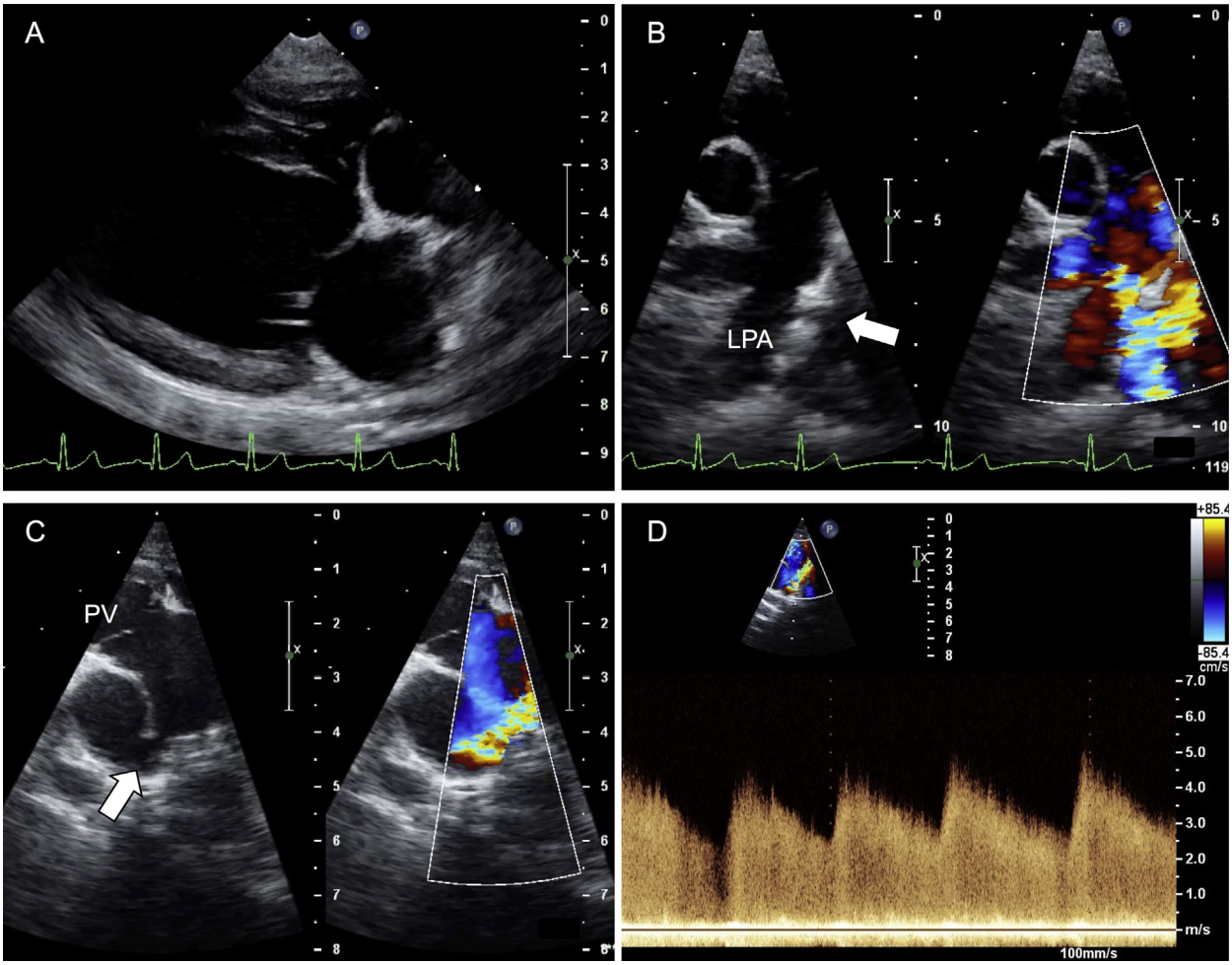

Figure 2 Transthoracic two-dimensional echocardiographic images. **(A)** Right parasternal long-axis four-chamber view. There is evidence of left ventricular and left atrial enlargement. **(B)** Right parasternal short-axis basilar view with color Doppler comparison. Note the atypical, almost perpendicular connecting angle of the continuously shunting vessel (*white arrow*) near the junction between the main and branch pulmonary arteries. **(C)** Left parasternal short-axis basilar view with color Doppler comparison. The same continuously shunting vessel (*white arrow*) appears to inset near the origin of the right pulmonary artery. **(D)** Continuous-wave Doppler indicating continuous left-to-right flow with an average peak velocity of 4.4 m/sec. *LPA*, Left pulmonary artery; *PV*, pulmonic valve.

must arise from the ascending aorta proximal to the origin of brachiocephalic arteries, and must terminate either in the dorsal aorta or in the pulmonary arteries via the persistently patent arterial duct."[10] The first component of the definition differentiates PFAA from a distal aortopulmonary window, which was already considered less likely in this case. The second component is present, and with regard to the third component of the definition, it is unclear if the insertion of the anomalous vessel to the right pulmonary artery is definitively through an arterial duct. The alternative to an arterial duct would be aortopulmonary collaterals; presumed PFAAs are often noted with pulmonary atresia, even though it is recognized that these collaterals typically arise from the descending aorta.[10] Nevertheless, given the lack of suitable alternative differentials for the anomalous vessel and absence of concurrent abnormalities that may compromise pulmonary blood flow, a PFAA remains a possible diagnosis in this dog.

In addition to the anomalous vessel, this case was unusual because of the concurrent finding of peripheral pulmonary artery stenosis that was not identified until angiography and computed tomography were performed. In humans, four types of peripheral pulmonary artery stenoses have been described: single stenosis involving the main pulmonary artery trunk or the right and left branches (type I), stenosis involving the bifurcation of the main pulmonary artery and extending into both branches (type II), multiple peripheral stenoses (type III), and a combination of main and peripheral stenoses (type IV).[11] The reported cases in small animals have either noted stenosis of the main pulmonary artery or multiple stenoses affecting both branch pulmonary arteries.[12-14] Many of these cases reported concurrent evidence of right ventricular concentric and/or eccentric hypertrophy and elevated right ventricular systolic pressures, but there was no evidence of right ventricular pressure or volume overload in this case. This may be because the stenosis was unilateral and the normal contralateral pulmonary arterial tree could accommodate the cardiac output without an increase in pressure. Interestingly, one case report in a cat found a PDA and juxtaductal coarctation of both pulmonary

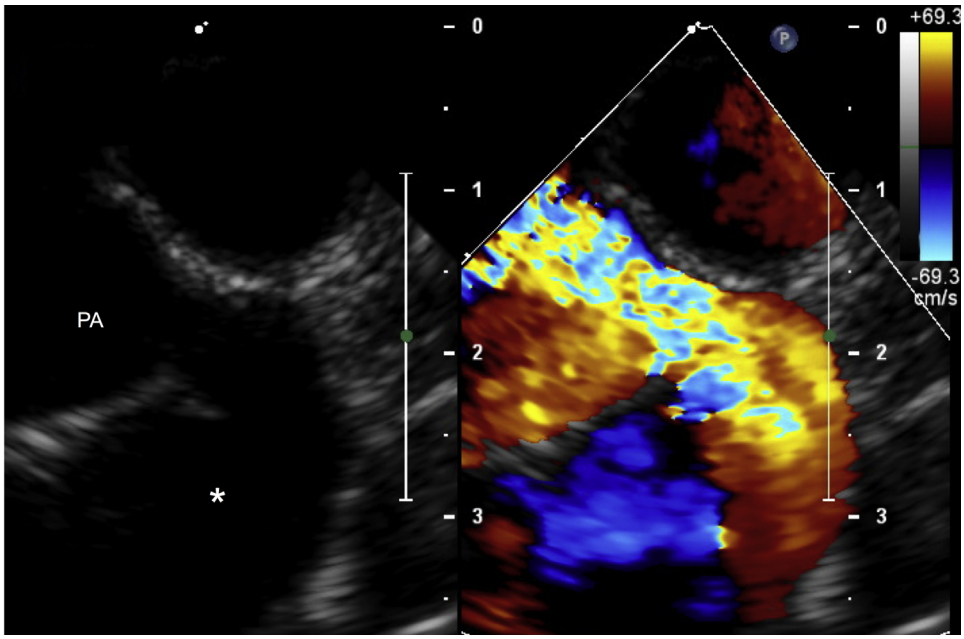

Figure 3 Transesophageal two-dimensional echocardiographic image with color Doppler comparison. The continuously shunting anomalous vessel (*asterisk*) is seen inserting into the ventral aspect of the pulmonary artery (PA).

arteries, with ectopic ductal tissue in the wall of the pulmonary arteries likely causing the constriction.[13] Given that the anomalous vessel in this case inserted into the right pulmonary artery, it is unlikely that it contributed to the left pulmonary artery stenosis. The pathogenesis of peripheral pulmonary artery stenosis remains incompletely understood and likely involves multiple factors and pathologic changes.

The predominant changes noted to the left heart indicate that the anomalous vessel was the more hemodynamically significant lesion compared with the unilateral pulmonary branch stenosis. The estimated pressure gradient across the anomalous vessel is smaller than would be expected if aortic and pulmonary artery pressures were normal, which may indicate a degree of pulmonary hypertension secondary to increased vascular resistance. Alternatively, the unilateral peripheral pulmonary artery stenosis may not have been significant enough to impart remodeling of the right heart but resulted in a degree of pressure elevation secondary to increased flow (diverted away from the left pulmonary artery). Last, some degree of misalignment of the Doppler interrogation beam (given the connecting angle of the anomalous vessel) may have led to underestimation of the gradient. Definitive determination of increased pulmonary vascular resistance would have necessitated direct measurements, but these were not obtained. Intravascular occlusion was attempted but not successful because of the inaccessibility of the anomalous vessel. Surgical ligation has been successful in other cases of anomalous vessels,

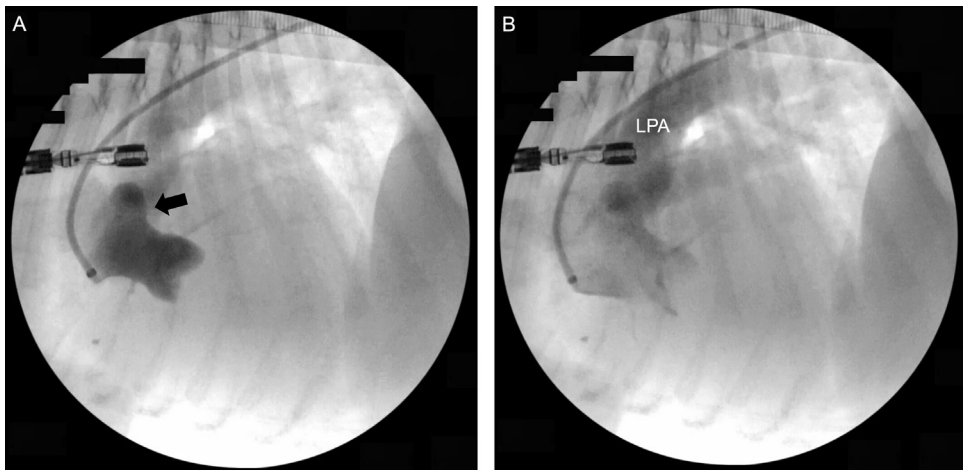

Figure 4 Aortic root angiography. **(A)** Upon initial injection, an anomalous vessel (*black arrow*) is seen originating from the dorsal aspect of the ascending aorta. Both coronary arteries are visualized and appear normal. **(B)** A notably enlarged left pulmonary artery (LPA) was highlighted shortly after.

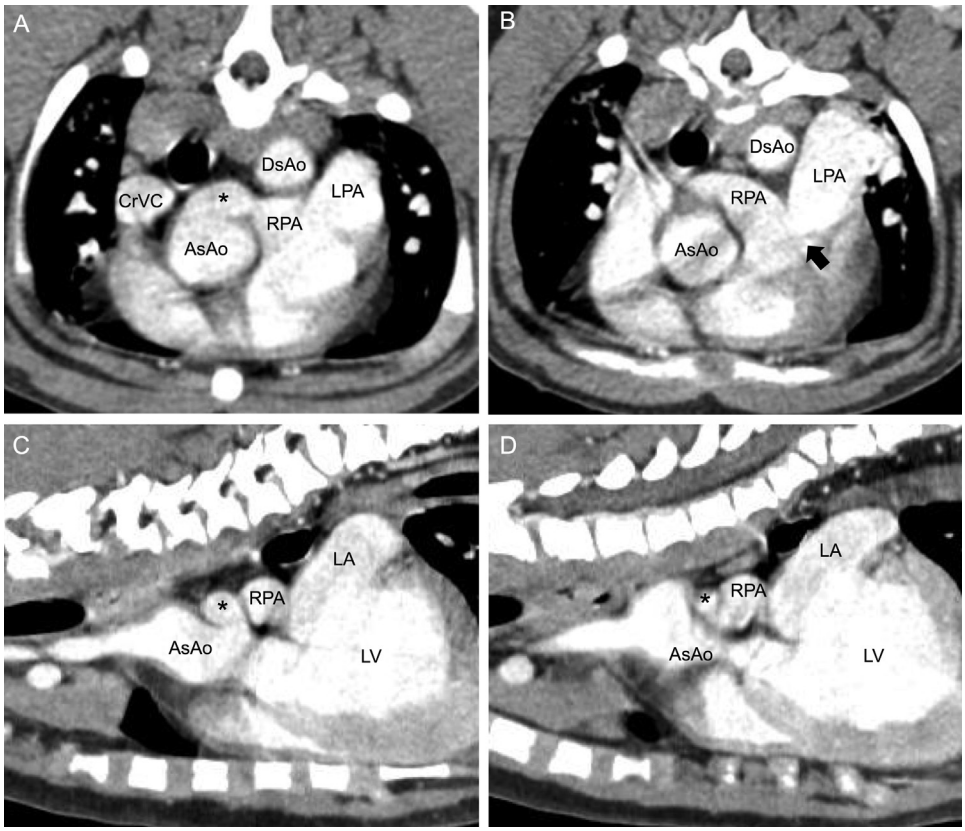

Figure 5 Computed tomographic angiography of the heart and major vessels. **(A)** Traverse plane at the level of the anomalous vessel (*asterisk*), which can be seen arising from the dorsal aspect of the ascending aorta (AsAo) and coursing toward the left to the right pulmonary artery (RPA). **(B)** Traverse plane highlighting the focal narrowing (*black arrow*) at the origin of the left pulmonary artery (LPA), indicating left peripheral pulmonary artery stenosis. Distally, the LPA is distended. In contrast, the RPA has a normal opening and size. **(C)** Sagittal plane centered at the origin of the anomalous vessel (*asterisk*). **(D)** Sagittal plane demonstrating the narrow insertion point of the anomalous vessel (*asterisk*) into the RPA. *CrCV*, Cranial vena cava; *DsAo*, descending aorta; *LA*, left atrium; *LV*, left ventricle.

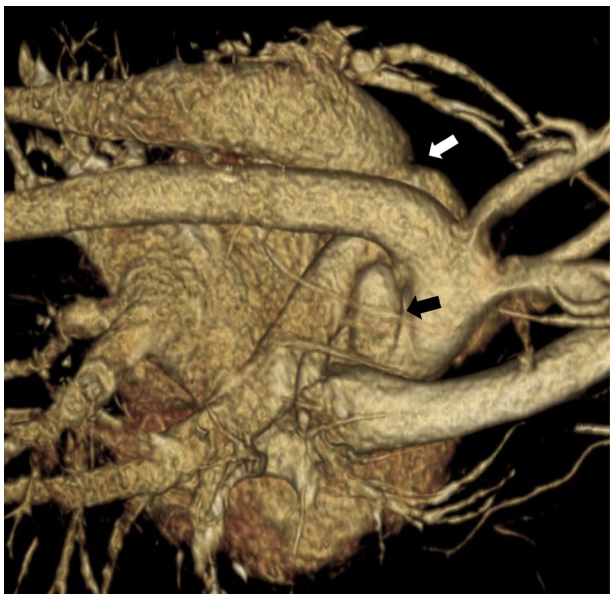

Figure 6 Computed tomographic three-dimensional reconstruction of the heart and major vessels. The anomalous vessel (*black arrow*) originates at the ascending aorta and extends to the right pulmonary artery. The site of the left peripheral pulmonary artery stenosis with poststenotic dilation is indicated by the *white arrow*.

although the owners declined because of increased risks for complications and financial limitations.[1,2] Although the prognosis for patients with this type of anomaly when untreated is unknown, it is presumed to be comparable with an untreated PDA given the similar pathophysiology. Treatment modalities for peripheral pulmonary artery stenosis include surgery, balloon angioplasty, and balloon-expandable intravascular stents, with stent placement becoming more commonplace in people because of improved outcomes compared with surgery or balloon angioplasty.[15] Treatment of the pulmonary artery stenosis was not pursued in this case given the unilateral nature of the stenosis. And if performed, surgical ligation of the anomalous vessel would likely have decreased the overcirculation to the pulmonary vasculature, further reducing the hemodynamic significance of the peripheral pulmonary artery stenosis.

CONCLUSION

This case illustrates that atypical appearances of continuous shunting on echocardiography may be further assessed with computed tomography and/or angiography for further characterization and identification of concurrent vascular anomalies that may have otherwise been masked or missed. Furthermore, although the findings were similar to those in previous reports, this may be the first case recognized as a possible PFAA in a dog.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2020.08.006.

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