# Partial Anomalous Pulmonary Venous Connection in 2 Miniature Schnauzers

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## Case 1

3-year-old, 6.0 kg, intact female Miniature A Schnauzer was presented to Azabu University for evaluation of right heart enlargement, incidentally noticed on survey thoracic radiographs. The dog was asymptomatic and no abnormalities were identified on physical examination. Radiographic evaluation of the thorax indicated right heart enlargement (vertebral heart score, 12.1). Two-dimensional echocardiography disclosed right atrial and right ventricular dilatation (Fig 1). An abnormal vascular structure connected to the right atrium at the heart base was observed on color Doppler echocardiography (Fig 2). No other structural heart disease or conditions that could result in right heart dilatation (eg, pulmonary hypertension, atrial septal defect [ASD], tricuspid valve regurgitation) were found. A complete blood count (CBC) and serum biochemistry profile were within normal limits. D-dimer concentration (reference range,  $<0.2 \ \mu g/mL$ ) was normal.

Computed tomography angiography (CTA) and cardiac catheterization were performed under general anesthesia, maintained by fentanyl constant rate infusion and isoflurane inhalation, to determine a definitive diagnosis. A 4 Fr multipurpose catheter was introduced from the right jugular vein through a catheter introducer placed by the Seldinger technique. Oxygen saturation (SaO<sub>2</sub>) of each site within the heart was measured while breathing room air. Mean SaO<sub>2</sub> of the cranial and caudal vena cava was 55.9%. SaO<sub>2</sub> in the right atrium varied from 81.8 to 99.2%, depending on the location of the catheter tip. SaO<sub>2</sub> at the right ventricle and pulmonary artery were 77.0 and 83.2%, respectively. Mean right atrial and pulmonary pressures were 1 and 11 mmHg, respectively.

The dog was placed in dorsal recumbency on a clinical 16-multi-detector-row computed tomography scanner. Iodinated contrast medium (2 mg/kg) was rapidly injected via the cephalic vein. Repetitive transverse

#### **Abbreviations:**

ASD	atrial septal defect
CTA	computed tomography angiography
MDCT	multi-detector computed tomography
PAPVC	partial anomalous pulmonary venous connection
SaO <sub>2</sub>	oxygen saturation

plane cine scans (120 kV, 99 mAs, 0.625 mm slice thickness, 0.6 s tube rotation time, 0.938 helical pitch) were acquired over the heart. Images were transferred to an image software system for further evaluation. Acquired images were analyzed using multiplanar reconstruction and volume rendering, and it was determined that the pulmonary vein of the right cranial lung lobe was connected to the right atrium. Therefore, a definitive diagnosis of partial anomalous pulmonary venous connection (PAPVC) was made (Fig 3).

## Case 2

An 8-month-old, 6.4 kg, intact female Miniature Schnauzer was presented for evaluation of right heart enlargement and a cardiac murmur. The dog was asymptomatic and no abnormality was identified on physical examination except a systolic heart murmur graded II of VI, and loudest at the left heart base. Radiographic evaluation of the thorax identified right heart enlargement (Fig 4; vertebral heart score, 11.0). On echocardiography, the same findings as observed in Case 1 were identified, which were right ventricular enlargement and an aberrant vascular structure at the heart base. Concurrently, doming of the pulmonic valve cusps during systole and mildly increased pulmonary blood flow (2.1 m/s) indicating mild pulmonic valvular stenosis. CBC and serum biochemistry profile were within normal limits. CTA and cardiac catheterization were performed using the same techniques as used in Case 1. SaO<sub>2</sub> of the caudal vena cava, right atrium, right ventricle, and pulmonary artery were 70.2, 78.7, 80.8, and 83.2%, respectively. Mean right atrial, systolic right ventricular, and systolic pulmonary pressures were 1, 31, and 20 mmHg, respectively. CTA disclosed that the pulmonary vein of the right cranial lung lobe was connected to the right atrium, as observed in Case 1 (Fig 5, 6).

Both cases were asymptomatic at the time of writing (34 and 29 months after diagnosis, respectively). There was no known familial relationship between the 2 dogs.

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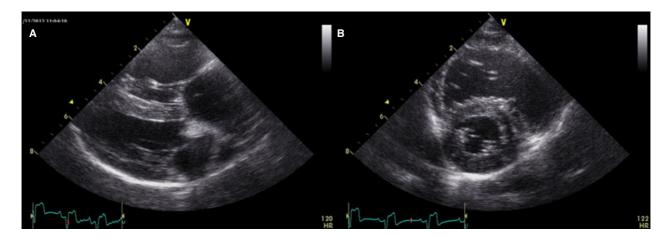
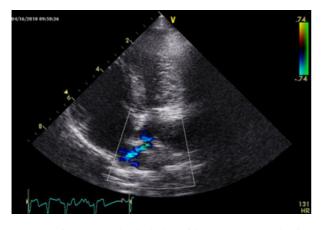


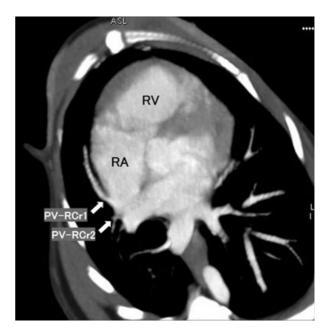
Fig 1. Right parasternal long axis (A) and short axis (B) views of the heart (Case 1). Right atrium and right ventricle were enlarged.



**Fig 2.** Left parasternal apical view of heart (Case 1). Color flow Doppler revealed an abnormal vascular structure connected to right atrium at heart base (arrows).

Right heart enlargement can be attributable to congenital heart disease, acquired conditions such as chronic valvular disease, or may be secondary to pulmonary disease. An intracardiac shunt, in particular ASD, is the most common congenital heart disease leading to enlargement of the right heart in dogs and humans. PAPVC is a congenital disease in which 1 or more (but not all) pulmonary veins are connected to the right atrium or cranial vena cava.<sup>1</sup> This anomaly also leads to right heart enlargement. Even if ASD is found in patients with right heart enlargement, when right ventricular enlargement is unexpectedly severe relative to a small ASD, PAPVC should be considered as a concurrent defect.<sup>2</sup>

Early atresia of the right or left portion of the common pulmonary vein, while primitive pulmonary-systemic venous connections are still present, will lead to PAPVC.<sup>3</sup> Studies in humans indicate that the most common form of PAPVC in children is right upper PV to the superior vena cava or to the right atrium. Anomalous connection of the left-sided pulmonary veins was much less common than anomalous connection of right-sided veins.<sup>1,4,5</sup> However, recent studies of adult



**Fig 3.** Transverse oblique image at level of right atrium (Case 1). Pulmonary vein from ventral part of right cranial lung lobe (PV-RCr1) and pulmonary vein from anterior part of right cranial lung lobe (PV-RCr2) flow together and drain into right atrium.

patients have identified that an anomalous left upper lobe vein is the most commonly affected vein in women, but it is infrequently associated with ASD.<sup>6,7</sup> In particular, in a case series of adults reported by Ho et al,<sup>7</sup> none of the patients had a suspected diagnosis of PAPVC at the time of imaging examination, suggesting that less severe cases may present during adulthood or be detected incidentally on imaging studies. In our series, both dogs had right-sided volume overload and anomalous pulmonary veins from the right cranial lung lobe. They were relatively young dogs, and presumably comparable to children. PAPVC is found in association with other cardiac defects, most commonly an ASD of the sinus venosus or secundum type in human,<sup>1,5</sup> although our cases did not have these defects.

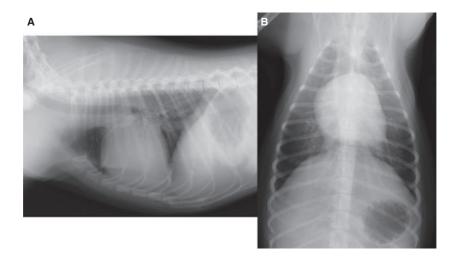
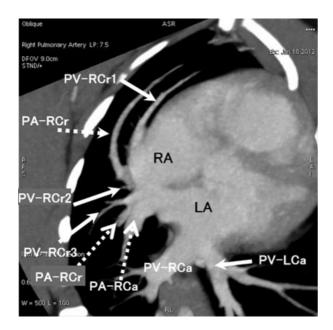
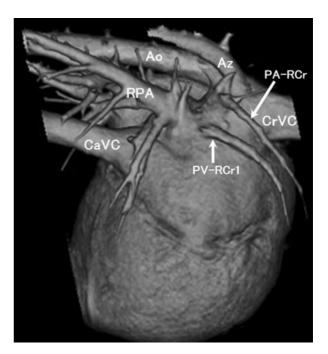


Fig 4. Thoracic radiographs of right lateral view (A) and dorsal-ventral view (B) in Case 2. Right heart enlargement was revealed.



**Fig 5.** Transverse oblique maximum intensity projection at the level of atrium (Case 2). Pulmonary vein from ventral part of right cranial lung lobe (PV-RCr1) and pulmonary vein from anterior part of right cranial lung lobe (PV-RCr2) flow together and drain into right atrium. Pulmonary vein from dorsal part of right cranial lung lobe (PV-RCr3) flows into left atrium. PA-RCr, pulmonary artery of right cranial lung lobe; PV-RCa, pulmonary vein of right caudal lung lobe; PV-LCa, pulmonary vein of left caudal lung lobe.

An anomalous pulmonary venous connection is an uncommon congenital disease in dogs.<sup>8,9</sup> In human medicine, PAPVC usually is diagnosed or suspected by echocardiography. Two-dimensional transesophageal echocardiography combined with color flow Doppler echocardiography has been reported as useful in diagnosing PAPVC.<sup>10,11</sup> However, proper technique is needed for identification of normal and abnormal pulmonary venous connections. MDCT angiography also



**Fig 6.** Right-sided view from a three-dimensional reconstruction of the heart and vasculature (Case 2). Pulmonary vein from ventral part of right cranial lung lobe (PV-RCr1) drains into right atrium. PA-RCr, pulmonary artery of right cranial lung lobe; CrVC, cranial vena cava; Az, azygos vein; Ao, aorta; RPA, right pulmonary artery; CaVC, caudal vena cava.

can reportedly provide more precise morphology of PAPVC. Cross-sectional imaging noninvasively and accurately evaluates the presence of anomalous veins. MDCT is ideal for this purpose, allowing recognition of anomalous vessels with high spatial resolution and wide anatomic coverage.

Computed tomography angiography has been used in dogs to evaluate not only the heart,<sup>12</sup> but also pancreas, liver, portosystemic shunts, anomalies of the vena cava and pulmonary vasculature.<sup>13–18</sup> Habing et al reported that MDCT allowed evaluation of pulmonary arteries at the subsegmental level in Beagle dogs.<sup>19</sup> They concluded that this technique had potential as a tool for the diagnosis of dogs with suspected pulmonary thromboembolism. The introduction of MDCT in veterinary medicine has markedly improved spatial and temporal resolution of pulmonary vessels. In the present cases, CTA enabled us to evaluate abnormally connected pulmonary veins in a small breed dog. CTA should be considered when pulmonary vascular abnormalities are suspected if a MDCT scanner is available.

Case 1 was previously described from our group as a case report,<sup>20</sup> and authors experienced case 2, the same breed and very similar clinical presentation to case 1. In addition, the authors examined 2 additional miniature schnauzers with right-sided volume overload that were suspected of PAPVC on the basis of echocardiography (unpublished data). Miniature Schnauzers may be predisposed to PAPVC because all 4 cases were of this breed. Additional studies are needed to determine the heredity of this congenital anomaly in dogs.

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