CONGENITAL HEART DISEASE IN DOGS AND HOW TO FIX IT

Surgical Management of Three Dogs with an Interventricular Communication and Atrioventricular Valve Abnormalities

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INTRODUCTION

Interatrial communications (IACs) are uncommon congenital heart defects in domestic dogs (Canis lupus familiaris). The oval fossa type (also known as secundum type IAC) and patent foramen ovale are most commonly described, followed by the primum type IAC in the form of an atrioventricular septal defect (AVSD).1,2 A small number of case reports have also described sinus venosus type IAC and unroofed coronary sinus (CS).3,4 While most IACs are small in size and do not cause any clinical signs during the dog’s lifetime, large IACs can result in significant right heart volume overload and ultimately congestive heart failure (CHF) and pulmonary hypertension.1,5 Closure of large IACs by open heart surgery or a transcatheter approach has previously been described.5,9 However, transcatheter closure often requires a discrete rim of atrial septum to stabilize the device and thus minimize the risk of embolization.5 An open heart approach is not restricted by this limitation but is more invasive, and reports have been limited to single dogs or experimentally created models.6,8,10,11

We describe three pet dogs that underwent open heart surgery to close a large IAC using expanded polytetrafluoroethylene (ePTFE). The summary of the types of IAC and basic cardiac dimensions are presented in Figure 1 and Table 1.

CASE PRESENTATIONS

Case 1

A 2-year-old female intact springer spaniel weighing 15.1 kg was examined for an incidental heart murmur. Physical examination showed a grade 4/6 left apical holosystolic murmur. The remaining physical examination was unremarkable.

Transthoracic echocardiography showed a large left-to-right shunting primum type IAC (1.4 cm) without a ventricular septal defect (VSD), consistent with a diagnosis of partial AVSD (Figure 1A). The dog displayed other characteristics of a partial AVSD, including the presence of atrioventricular (AV) valvar leaflets guarding the common AV junction, with the zone of apposition from the two bridging leaflets of the left AV valve pointing toward the septum (Figure 2). There were two AV valvar regurgitant jets— moderate-to-severe regurgitation jet from the left AV valve and a small regurgitation jet from the right AV valve, both directing toward the right atrium (RA). The estimated pulmonary arterial systolic pressure using the right AV valvar regurgitant velocity was 58 mm Hg. The RA and right ventricle were severely enlarged (Table 1).

While the dog was not showing any clinical signs, the risk of developing clinical signs from the large IAC was deemed high. After a careful discussion with the owner, the decision to pursue the surgical closure of the IAC was made.

General anesthesia and cardiopulmonary bypass (CPB) were established as previously described.11 A detailed surgical description is provided in Video 1. At surgery, the AV valves were inspected and the two bridging leaflets of the left AV valve were sutured together before closing the IAC using a 0.4 mm thick patch of ePTFE (Gore-Tex Cardiovascular Patch, W. L. Gore & Associates, Inc., Flagstaff, AZ). A distinct remnant or stump of an atrial septum was seen adjacent to the Eustachian valve and the CS ostium so that the patch could be secured while conserving the normal relationship between the CS and RA. A small patent foramen ovale that was not seen during presurgical imaging was also identified and closed. The postop transesophageal echocardiography showed mild residual left AV valvar regurgitation.

On discontinuation of CPB, the dog developed sinus arrest with a low ventricular escape rate (20 beats per minute), likely due to an inappropriately slow sinus nodal recovery. This was managed using temporary pacing for 24 hours until sinus rhythm returned. However, frequent atrial premature complexes (APCs) became apparent, and these later progressed into persistent atrial flutter (AFL) on day 2 following the surgery. This was managed using oral sotalol and transthoracic electrical cardioversion on day 3 without complication. The dog was discharged 2 weeks postop with aspirin and clopidogrel for 3 months (our standard antiplatelet protocol).

A follow-up assessment 1 month postop showed that the dog remained clinically asymptomatic. A repeat echocardiogram showed substantial reverse remodeling of the right heart (Table 1). There was no residual shunting in the IAC, and the left AV regurgitation was trivial. Subsequent follow-up 4 years postop by the referring veterinarian showed that the dog continued to do well clinically with normal exercise levels and was not receiving any medications.
Case 2
A 2-year-old female intact Labrador weighing 23.6 kg was assessed for an incidental grade 6/6 right-sided holosystolic murmur. The heart rate was 140 beats per minute with strong and synchronous femoral pulses. The jugular veins were distended. Other physical examination findings were unremarkable.

Transthoracic echocardiography showed a large (3.4 cm) left-to-right shunting IAC (Figure 1B). There was some remnant of the interatrial septum around the AV junction, roof of the atrium, and the Eustachian valve, providing some surgical landmarks (Figure 3). An imaginary line from the tissue at the roof of the atrium to the AV junction appeared to divide the left atrium (LA) and RA with appropriate venoatrial connection. In addition to the IAC, the dog was diagnosed with tricuspid valve dysplasia, with long and tethered leaflets attached to thin papillary muscles (Figure 4, Video 2). Color Doppler showed severe tricuspid regurgitation (Video 3). Paradoxic septal flattening was present, consistent with right ventricular volume overload. Finally, the CS was distended, suggestive of an increased right atrial pressure and/or persistent left cranial vena cava (PLCVC). There were no other abnormalities identified.

**Figure 1** Schematic diagrams for the corresponding echocardiographic images of three dogs with large IACs. The right parasternal long-axis four-chamber view at both pre- and postsurgery is displayed for each dog. The dotted line outlines the interatrial communication in each image. The first dog (A) had an ostium primum type IAC (red line), consistent with partial AVSD. The second dog (B) had a large oval fossa type IAC (blue line) and tricuspid valve dysplasia with severe tethering of the septal tricuspid valve leaflet (arrow). The third dog (C) had a large IAC, with a feature of ostium primum type IAC (orange line), which together with a small restrictive VSD (green line) was consistent with intermediate AVSD. The IAC was closed in all three dogs using a patch of 0.4 mm ePTFE. Ao, Aorta; CdVC, caudal vena cava; CrVC, cranial vena cava; LV, left ventricle; PV, pulmonary vein; RAA, right atrial appendage; RV, right ventricle; TV, tricuspid valve.
As with the previous case, a decision was made to proceed with a surgical intervention to mitigate the risk of CHF and pulmonary hypertension. A presurgical cardiac computed tomography (CT; non–electrocardiogram [ECG] gated) was performed to further evaluate the venoatrial connections and the presence of PLCVC.

The CT confirmed the presence of a PLCVC (Figure 5). The PLCVC was draining into the CS between the left atrial appendage and the left pulmonary veins. The right cranial vena cava was unremarkable. The pulmonary veins were also unremarkable and were draining into the morphological LA.

The surgical description is provided in Videos 4 and 5. The tricuspid valve was inspected, and the tethering of the septal leaflet by numerous short primary and secondary chordae tendineae was confirmed. The mural leaflet had subjectively normal motion despite abnormal chord/papillary muscle attachments. A similar approach was used for the tricuspid valve repair in a recent publication. The septal leaflet was released from the short primary and secondary chordae tendineae by sharp dissection. The septal leaflet was resuspended ddae tendinae by sharp dissection. The septal leaflet was resuspended.

Similar to case 1, clopidogrel and aspirin were also given for 3 months. Repeat cardiac assessment 1 month, 4 months, and 12 months postsurgery showed evidence of reverse remodeling (Table 1) and some reduction in tricuspid regurgitation, which remained severe (Videos 6 and 7). The dog’s quality of life was perceived to be excellent by the owner 3 years after the surgery.

Case 3

A 1.5-year-old female intact fox terrier weighing 7.6 kg was presented for evaluation of a heart murmur. Physical examination confirmed a grade 5/6 right-sided holosystolic murmur. The femoral pulses were strong and synchronous, and physical examination was otherwise normal.

Transthoracic echocardiography revealed a large left-to-right shunting ostium primum type I AIC (2.7 cm) accompanied by a small inlet-type VSD (4 mm) with left-to-right shunting, consistent with an intermediate AVSD (Figure 1C). Most of the atrial septum was missing, although a small 5 mm strand of tissue was seen on the roof of the common atrium. Similar to case 2, an imaginary line across this strand of tissue to the ventricular septum appeared to divide the morphological LA and RA with appropriate venoatrial connections. There were four AV valvar leaflets, with the bridging leaflets of the left AV valve creating a zone of apposition pointing toward the septum (Figure 6). There was mild left AV valvar regurgitation and severe right AV valve regurgitation. The right ventricle was severely enlarged with paradoxical septal motion consistent with right ventricular volume overload. The pulmonary artery was severely dilated.

Surgical repair of the AVSD was scheduled. Presurgical ECG-gated cardiac CT and three-dimensional (3D) transesophageal echocardiography confirmed the transthoracic echocardiographic findings and better showed the cardiac anatomy including other features of intermediate AVSD (Figures 6 and 7, Video 8).

The surgery (Video 9) was very similar to case 1, with the difference being closure of the inlet-type VSD. The VSD closure was not recorded in Video 9. Briefly, the VSD was closed using a single mattress suture from the left side, with pledget in on the ventricular side. The same suture was then used to suture the patch from the atrial side.

### Table 1 Basic cardiac dimensions at baseline and 1-month after corrective surgery (postop)

| Two dimensions | 95% RI | Partial AVSD | Fossa oval type IAC, tricuspid valve dysplasia, PLCVC | Intermediate AVSD |
|----------------|-------|--------------|---------------------------------------------------|------------------|
| Dog 1          |       | Baseline     | Postop                                                  | Baseline         |
| RA <sub>min</sub> <sup>2D, Apex</sup>, mm | 6.12-9.02 | 16.34 | 14.01 | 16.93 | 14.01 | 17.41 | 14.06 |
| RVd<sub>min</sub> <sup>2D, Apex</sup>, mm | 5.27-9.35 | 15.7 | 14.1 | 13.92 | 10.96 | 16.44 | 12.29 |
| LA<sub>2D, Lx</sub>, cm | 1.19-1.56 | 1.26 | 1.50 | 1.17 | 1.37 | 1.05 | 1.52 |
| LVID<sub>d2D, Lx</sub>, cm | 1.15-1.55 | 1.49 | 1.48 | 1.05 | 1.16 | 1.04 | 1.32 |
| LVID<sub>s2D, Lx</sub>, cm | 0.68-1.09 | 0.97 | 1.03 | 0.79 | 0.68 | 0.55 | 0.56 |
| LV FS% | 19.1-41.7 | 34.9 | 30.4 | 24.8 | 41.4 | 47.1 | 57.6 |

The measurements were taken as previously described over three consecutive cardiac cycles and indexed to body size using the equation: constant = measured dimension/body weight (kg) <sup>scaling exponent</sup>. The 95% reference intervals (RIs; 2.5% centile to 97.5% centile) for each indexed variable are shown. All dogs showed marked reduction in right heart dimensions and some increase in left heart dimensions at the 1-month recheck postoperatively.
Figure 2  Echocardiographic and surgical images for case 1. The AV valves are shown in the right parasternal short-axis view in pre- and postop. The direction of the zone of apposition is marked with an orange arrow. IBL, Inferior (caudal) bridging leaflet; LML, left mural leaflet; SBL, superior (cranial) bridging leaflet.

Figure 3  Three-dimensional echocardiography of the IAC showing the outline of the atrial septum. The red arrow points at the Eustachian valve.
Although an abnormal right AV valve was confirmed by direct inspection, a saline leak test revealed subjectively good valve leaflet coaptation. Based on the assumption that a proportion of the valve regurgitation was secondary to right heart overload, and concern that an attempt to repair the right AV valve might worsen valve function, the right AV valve was not repaired. Intraoperative transesophageal Doppler echocardiography showed residual mild left AV regurgitation and minimal residual flow through the VSD.

As in the case for dogs 1 and 2, this dog required temporary pacing until day 3 postop. Frequent APCs were also seen with a single self-limiting paroxysmal AFL seen a day following the surgery, but medical management was not required. The dog was discharged on day 7 postoperatively on a 3-month course of clopidogrel and aspirin. A recheck 1 month later showed improved cardiac dimensions (Table 1). The residual flow through the VSD was no longer present. Further follow-ups at 4 and 9 months postsurgery showed further reverse remodeling of the heart and mild persistent left AV valvular regurgitation and only mild right AV valvular regurgitation. The owner perceived that the dog had an excellent quality of life at home 1 year after the surgery.

**DISCUSSION**

We have described three dogs that underwent successful surgical treatment of their large IACs and associated or concurrent AV valve abnormalities under CPB. A small inlet-type VSD (as part of the AVSD disease process) in one dog was also closed. None of these dogs were showing any clinical signs. However, many dogs with large IACs remain asymptomatic until they are older (i.e., 4 years old), and the risk of developing CHF or pulmonary hypertension was considered high for each animal in this report. The reverse remodeling following surgical intervention confirmed our justification for surgical correction. Successful surgical management of large IACs with concurrent congenital AV valve abnormalities, which depends heavily

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**Figure 4** Preop and postop right parasternal long-axis four-chamber views of dog 2. Note the severe right heart enlargement with apical displacement of the septal tricuspid valve leaflet hinge point by 1 cm due to severe tethering (green line). Postsurgical image shows an improved motion of the septal tricuspid valve leaflet and an artificial atrial septum. The red arrow shows the Eustachian valve as also shown in Figure 3.

**Figure 5** Three-dimensional reconstruction of the CT performed preoperatively, viewing from the left side (A) and the dorsal aspect (B). Note the PLCVC (outlined in a dotted line). The PLCVC drained into the CS between the pulmonary veins and left atrial appendage. The contrast intensity was not consistent in the PLCVC, making it irregular on 3D reconstruction, but the walls were parallel and there was no stricture. Ao, Aorta; LV, left ventricle; PA, pulmonary artery; RV, right ventricle.
on accurate morphological characterization of the defects, has rarely been described in pet dogs. In the series of dogs reported here, we have presented the presurgical imaging that was essential to the development of each surgical plan, along with the details of the surgical procedures performed. At the time of writing, all of these dogs are enjoying a normal quality of life, according to their owners, despite continued mild AV valve dysfunction.

Several factors influenced the decision whether to place the patch on the left or right side of the CS. In AVSD, the AV node is located closer to the CS in the triangle of Koch, and extending the patch rightward to the CS is commonly suggested to avoid the conduction system. However, sutting the patch on the left side of CS has also been suggested, and it provides some benefits if it can be safely performed. First, CS drainage in the RA is physiologically appropriate.
Second, in cases where the left atrial pressure is increased due to significant left heart disease, there can be a deleterious effect on coronary venous return. While we were aware of the risk of damaging the AV node in each case, the surgeon could identify a clear remnant of an interatrial septum in the cranial and caudal borders of the inner atrial wall in both AVSD cases (cases 1 and 3). Based on this, we assessed that careful placement of superficial suture bites would be low risk as described in pediatric cardiac surgery. Neither dog developed an AV block postoperatively. In contrast, case 2 had a large IAC that lacked a clear surgical landmark in the caudal border of the IAC. Therefore, the surgeon chose to extend the patch rightward to the CS and avoid any risk of damaging the AV node. This decision-making was possible as the presurgery CT scan showed both PLCVC and a right cranial vena cava, and thus the PLCVC could be ligated if the CS had to be reassigned to the LA.

As there is currently no consensus or guidelines on the classification of IACs in dogs, this report followed the recommended anglicized nomenclature from the International Paediatric and Congenital Cardiac Code (IPCCC) with some modifications based on Nomina Anatomica Veterinaria–approved veterinary terminology. We have found that consistent use of this nomenclature and the use of multimodal imaging are particularly useful for avoiding ambiguous descriptions during surgical planning for the dogs described here. Accurate classification of large IACs such as those seen in dogs 2 and 3 was difficult, as the possibility of multiple IACs to create a single large IAC could not be ruled out. Two dogs underwent multimodal imaging, which allowed verification of the transthoracic echocardiographic findings and accurate assessment of the venoatrial connections in relation to the IAC. Furthermore, 3D transesophageal echocardiography allowed a careful
examination of the AV valve, and this assessment proved to be consistent with the intraoperative assessment. The multidisciplinary approach, along with good knowledge of both normal and abnormal cardiac morphology and coupled with standardized descriptors, is essential for successful management of dogs with such complex congenital heart disease.

The left AV valve in the two dogs with AVSD appeared to be “cleft” because the two bridging leaflets create a zone of apposition toward the septum. As previously stated, we have used the nomenclature from the IPCCC and decided against using the term “cleft” or “mitral valve” in AVSD, as this is a common valve and not a true cleft. In people, true MV cleft is associated with an abnormal MV attachment to the left ventricular outflow tract, resulting in the cleft pointing toward the left ventricular outflow tract. A similar finding has been described in a veterinary case report.

All three dogs developed arrhythmias postoperatively. The types of arrhythmias were sinus arrest post-CPB and supraventricular arrhythmias (frequent APCs and paroxysmal or persistent AFL). These were managed using temporary cardiac pacing, oral antiarrhythmics, and electrical cardioversion, without the need for long-term management. Similar arrhythmias have been described in people undergoing cardiac surgery, with possible etiologies that include acute hemodynamic changes, ischemia, infarction, hypertension, trauma, inflammation, electrolyte derangement, and cardiac medications. Specific to the dogs reported here, acute hemodynamic changes, trauma from the tissue handling, inflammation, or infarction were considered the most likely given the transient nature of these arrhythmias. Unintentional suturing of the conduction system and damage to the sinoatrial node while cannulating the cranial vena cava were also possible causes but were considered less likely based on our experience of comparing different cannulation techniques.

CONCLUSION

Interatrial communication and AV valve abnormalities can be successfully managed surgically in dogs. Careful morphological description, particularly with multimodal imaging, greatly assists the surgical planning.

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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2021.05.002.

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