Gerbode defect in a dog

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Abstract: An intracardiac communication between the left ventricle and right atrium (Gerbode defect) was diagnosed in a 9-year Yorkshire Terrier with a history of chronic exercise intolerance. The history, clinical examination, and diagnostic imaging confirmed the diagnosis and did not reveal evidence of trauma or endocarditis which could lead to this special type of left-to-right shunting. A Gerbode defect is a very rare finding in human beings and animals. In the veterinary literature all reports about this condition were related to thoracic trauma or valvular infection. According to the authors, this would be the first clinical case of congenital Gerbode defect in a dog.

Keywords: canine; cushion defect; echocardiography; ventricular septal defect

Gerbode defect is a rare type of ventricular septal defect in which there is communication between the left ventricle and right atrium. In human medicine, Gerbode defects may account for about 0.08% of all congenital heart diseases (Ramirez et al. 2003; Vijayalakshmi et al. 2013). In general, a ventricular septal defect can be located along the wall of the interventricular septum, but they are more commonly observed in the upper and posterior part known as the membranous interventricular septum, which separates the aortic vestibule from the lower part of the right atrium and the upper part of the right ventricle. Furthermore, the membranous part can be divided by the septal leaflet of the tricuspid valve into the atrioventricular and interventricular parts (Figure 1A). Type I (supravalvular or atrioventricular) defects occur in the atrioventricular part of the membranous septum resulting in direct communication of the left ventricle to the right atrium (Figure 1B). Type II (infravalvular or interventricular) defects are indicative of a true ventricular septal defect associated with other congenital changes in the septal leaflet of the tricuspid valve (such as perforations or clefts, fenestration, aneurysmal transformation or intergrowths of valve leaflets to the edges of the defect) (Figures 1B–1D) (Ramirez et al. 2003; Peddle et al. 2008; Karaci et al. 2012). In these cases, shunting of blood is observed not only from the left ventricle to the right ventricle but also to the right atrium, which is achieved by crossing through the abnormalities in the septal tricuspid leaflet. In some instances, a unique defect involves both supravalvular and infravalvular (type III) abnormalities (Figure 1D) (Peddle et al. 2008; Sinisalo et al. 2011). Human cases of Gerbode defect can be recognised congenitally or secondary to cardiac surgery (e.g. valvular transplantation), septic endocarditis, thoracic trauma and myocardial infarction (Ramirez et al. 2003; Peddle et al. 2008; Hezzell et al. 2012).

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never suffered from any other condition with the exception of unilateral patellar luxation that was successfully corrected three years earlier. The referring veterinarian found a murmur and prescribed benazepril (0.25 mg/kg p.o., s.i.d.). The dog was up-to-date regarding vaccinations and deworming. On presentation, the dog was mentally alert with pink and moist mucous membranes. There was a mild degree of dental tartar. Auscultation revealed tachycardia (160 bpm) with regular femoral pulses. Additionally, a sternal holosystolic heart murmur was detected at the left hemithorax. The respiratory rate was 28 rpm with normal bronchovesicular sounds. At the time, differential diagnoses pointed to degenerative mitral valve disease or endocarditis. Diagnostic work-up included electrocardiographic examinations such as an electrocardiogram (ECG), thoracic radiographs, blood pressure measurement and echocardiography.

A 6-lead ECG (Figure 2) revealed sinus arrhythmia with a heart rate of approximately 140 bpm and the presence of T waves and occasional tall P waves. Blood pressure obtained from the dorsal (pedal) metatarsal artery using a Doppler ultrasonic system revealed a normal systolic blood pressure of 130 mm Hg. Thoracic radiographs demonstrated a mildly enlarged cardiac silhouette (vertebral heart

**Case description**

A nine-year spayed female Yorkshire Terrier dog was referred to the Small Animal Clinic at the University of Veterinary and Pharmaceutical Sciences Brno, Czech Republic, with a history of exercise intolerance for several weeks. The dog had
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left ventricular outflow tract that crossed the peri-membranous portion of the septum towards the right ventricle as well as towards the right atrium proximally to the level of the septal tricuspid leaflet (Figure 4). A modified right parasternal 5-chamber view was used to further investigate shunting from left ventricle-to-right atrium, confirming a connection between the atrioventricular part of the membranous interventricular septum and the right atrium (Figure 5). Spectral continual Doppler estimated a gradient through the left-to-right shunting of 3.3 m/s (approximately 44 mm Hg) (Figure 6). Also, mild degenerative changes in the mitral and tricuspid valves were recorded from left apical views. The pulmonary-systemic flow ratio (Qp/Qs) scale = 11) without pulmonary venous congestion. The trachea presented different degrees of lumen irregularities and there was a dominant bronchial lung pattern (Figure 3). Complete blood count and serum chemistries were unremarkable.

Standard echocardiography showed thickening of the caudal leaflet of the mitral valve and a mildly hyperechogenic and elongated valve apparatus of the cranial tricuspid leaflet. There was no prolapse of any of the atrioventricular valves. Systolic left atrial size at M-mode was 22.3 mm and the left atrium/aorta ratio was 1.42. The other 2D and M-mode parameters were within the normal range (Boon 2011). Colour flow mapping displayed turbulent flow originating within the left ventricular outflow tract that crossed the peri-membranous portion of the septum towards the right ventricle as well as towards the right atrium proximally to the level of the septal tricuspid leaflet (Figure 4). A modified right parasternal 5-chamber view was used to further investigate shunting from left ventricle-to-right atrium, confirming a connection between the atrioventricular part of the membranous interventricular septum and the right atrium (Figure 5). Spectral continual Doppler estimated a gradient through the left-to-right shunting of 3.3 m/s (approximately 44 mm Hg) (Figure 6). Also, mild degenerative changes in the mitral and tricuspid valves were recorded from left apical views. The pulmonary-systemic flow ratio (Qp/Qs)

Figure 2. ECG tracing revealed sinus arrhythmia with a heart rate of approximately 140 bpm and the presence of T waves (black arrows) and occasional tall P waves (asterisks), which may be a change suggesting right atrial overload. Wandering sinus pacemaker can also be seen. Paper speed 50 mm/s

Figure 3. (A) Laterolateral view. Vertebral heart scale measures approximately 11 vertebral bodies. The tracheal lumen can be distinguished as irregular based on its thoracic length. Some spondylosis could also be seen at the 4th, 5th and 6th thoracic vertebral bodies. A lung bronchial pattern also can be seen at caudal lung lobes (arrows). (B) Dorsoventral view. Mildly rounded heart silhouette. There is an apparent mild collapse of the left main bronchus (black circle). Also, the lung bronchial pattern is evident on both sides but is more marked on the right middle lung lobe (white arrows)
was determined at 1.2 (< 2:1 is haemodynamically insignificant) (Boon 2011). Based on the results of the cardiological examination, a supravalvular (type I) Gerbode defect was diagnosed. The owner declined other more invasive investigations such as contrast studies or catheterisation. The patient was sent home with the same therapy and after six months the follow-up did not show any worsening in clinical or echocardiographic situations.

Figure 4. (A) Modified right parasternal long axis view at the level of left ventricular outflow tract showing the presence of a ventricular septal defect (arrow) between the left ventricle and right atrium. (B) Simultaneous colour flow mapping (CFM) displaying a left-to-right shunting through the defect. (C) Right parasternal short axis view at the level of the right ventricular outflow tract and aorta. CFM demonstrates abnormal blood shunting through the septal defect from the left ventricle to both the right atrium and right ventricle

AO = aorta; LVOT = left ventricular outflow tract; RA = right atrium; RV = right ventricle; RVOT = right ventricular outflow tract; TV = tricuspid valve

DISCUSSION AND CONCLUSIONS

The first description of Gerbode defect was reported in humans in 1857 (Meyer 1857). Most Gerbode-type defects in humans are congenital and have been reported in association with other congenital heart defects, including subaortic stenosis, atrial septal defect and other vascular anomalies (Vijayalakshmi et al. 2013). Interestingly, human patients with congenital Gerbode defects (and other congenital diseases) are at a higher risk of developing infective endocarditis. The risk of infective endocarditis is closely related to abnormal intracardiac shunts and high-velocity or turbulent flows (Karaci et al. 2012). In the dog of this report, other congenital or acquired diseases were not observed with the exception of mild degenerative changes in the mitral valve (ACVIM classification B1), a condition that would be more typical of such a patient due to the age and breed.

An acquired Gerbode defect in humans may be secondary to cardiosurgery, endocarditis, thoracic trauma or myocardial infarction. It was first reported in the veterinary literature as a necropsy finding by Ramirez et al. (2003). Two of the four reports on Gerbode defects that have been so far described in canine patients were secondary to bacterial endocarditis (Ramirez et al. 2003; Peddle et al. 2008), and the other two cases were found secondary to thoracic trauma (in one case associated with atrial septal defect, probably also traumatic).

Figure 5. Modified right parasternal long axis view. Colour flow mapping (CFM) reveals a systolic flow from left ventricular outflow tract (LVOT)-to-right atrium (RA) above the tricuspid valve

LA = left atrium; LV = left ventricle; MV = mitral valve; RV = right ventricle; TV = tricuspid valve
(Hezzell et al. 2011; Cunningham et al. 2013). No traumatic incident or any cardiac infection was described in the history, suspected in the clinical exam nor confirmed with other diagnostic tests in our patient. We assume that the origin in this case was congenital; however, and according to human reports, most supravalvular defects are acquired conditions (Ramirez et al. 2003). This may be due in part to the associated lesions seen on the septal leaflet of the tricuspid valve.

In general, dogs with ventricular septal defect may develop clinical signs normally seen in young dogs (one to two years of age) but can present at an older age with symptoms of generalised weakness, cough, exercise intolerance, syncope, abdominal swelling associated with ascites and pale mucous membranes. Other patients may not present signs at all, and ventricular septal defects are found as incidental findings during other examinations. Dogs with the Gerbode defect may show acute symptomatology due to a destructive perforating endocarditis or blunt trauma leading to rupture of the membranous interventricular septum. Signs of systemic infection (due to endocarditis-like fever, lethargy, anorexia or weight loss), arrhythmia or signs associated with trauma (laboured respiration, bleeding or lameness) may precede signs of heart failure (Hezzell et al. 2011; Cunningham et al. 2013). The patient of this report was a geriatric dog with vague symptomatology of exercise intolerance, which does not reflect an acute process; however, past valvular infections could have occurred and then subsequently healed. ECG abnormalities were numerous. We found ECG findings of right atrial overload due to the presence of peaked tall P waves in combination with T waves (Tilley 1992). These findings may suggest long-term right-side overload as seen in other reports (Cunningham et al. 2013). Other described ECG findings are VPCs (ventricular premature complexes) (Hezzell et al. 2011) and varying degrees of atrioventricular blocks (Cunningham et al. 2013) or left or right bundle branch blocks (Vijayalakshmi et al. 2013). This may be due to the proximity of ventricular septal defects to the atrioventricular conduction tissue in the membranous interventricular septum or could be secondary to direct damage to the conduction system in the atrioventricular node as seen in canine and human patients (Peddle et al. 2008; Sinisalo et al. 2011). Long-term shunting may lead to biventricular volume overload and ventricle enlargement (Silbiger et al. 2009). To date, the patient is doing well on medical management alone; her estimated Qp/Qs is an indication that pulmonary flow exceeds systemic flow and defines a net left-to-right shunt that remains below the threshold for any other intervention (1.5–2 : 1 is considered indication for intervention). Treatment is usually conservative, and the goals are to prolong the onset of overt heart disease and management of the secondary congestive heart failure; however, surgical correction can be performed at specialty referral centres. In humans, the condition is usually repaired surgically, mainly by using either percutaneous Amplatzer ductal or septal occluder devices. The first successful closure of
such a defect was reported by Kirby (using hypothermia and inflow occlusion) at the Hospital of the University of Pennsylvania in 1956 (Kirby et al. 1957). The first successful series of operations on patients with a left ventricular-to-right atrial shunt and a complete description of this condition was reported by Gerbode et al. (1958). According to the veterinary literature, the proximity to the aortic valve and the ratio device size and patient size makes it difficult to deploy equipment in very small patients. A more invasive approach in this patient was not considered due to the restrictive nature of the defect, lack of clinical signs and the size of the dog.

REFERENCES


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