Double-chambered right ventricle (DCRV) is a congenital heart defect characterized by an anomalous fibromuscular band or membrane at the junction of the inflow and outflow portions of the right ventricle. The anomaly causes varying degrees of obstruction to flow and pressure overload within the right ventricle. Hypertrophy develops only in the inflow portion of the right ventricle, giving the ventricle a “double-chambered” appearance.

Double-chambered right ventricle has been reported in 3 dogs. Despite these reports, DCRV is not widely recognized as a distinct cardiac anomaly in dogs. Further, this anomaly may have emerging importance and prevalence as a congenital heart defect in certain breeds of dogs. This report describes the clinical findings and surgical correction of DCRV; the intermediate-term outcomes of 7 dogs that underwent correction for DCRV are reported.

Methods

Diagnostic evaluation—Diagnostic evaluation for dogs with suspected congenital heart defects consisted of physical examination, blood and urine analyses, thoracic radiography, standard 6-lead ECG, Doppler-measured systolic blood pressure, and 2-dimensional and Doppler echocardiography. Diagnostic criteria for DCRV consisted of demonstration of a discrete obstruction separating the inflow and outflow portions of the right ventricle, concentric hypertrophy of inflow portion of the right ventricle, and a normal thin-walled appearance of the right ventricular outflow tract (RVOT). Standard 2-dimensional echocardiographic imaging planes were used to reveal the presence of right ventricular hypertrophy. Most often, standard 2-dimensional echocardiographic imaging planes were not optimal to view the obstruction. An oblique right parasternal transverse view of the base of the heart and left ventricle and the left cranial transverse view optimizing the tricuspid valve were used to see the obstruction proximal to the pulmonic valve (Fig 1). The peak flow velocity (V) across the area of obstruction in the right ventricle was measured by use of continuous-wave Doppler imaging. The maximum instantaneous pressure gradient (PG) across the defect was calculated from V by use of the simplified Bernoulli equation (PG = 4V²). Color flow Doppler echocardiography was used to direct alignment of the Doppler cur.

Figure 1—Echocardiogram from a dog with double-chambered right ventricle (DCRV). A—Right parasternal transverse view optimizing the heart base was obtained with the dog in right lateral recumbency. The obstruction can be seen proximal to the pulmonic valve. B—Right parasternal transverse view optimizing the left ventricle. The obstruction can be seen distal to the tricuspid valve. C—Left cranial transverse view (obliqued to optimize the tricuspid valve) obtained with the patient in left lateral recumbency. The obstruction is seen above the tricuspid valve. OB = Obstruction. RA = Right atrium. RV = Right ventricle. PV = Pulmonic valve. TV = Tricuspid valve. RVOT = Right ventricular outflow tract. LV = Left ventricle.
Anesthesia, cardiopulmonary bypass, and cardiac repair—Dogs were medicated with atropine (0.03 mg/kg [0.014 mg/lb], SC) or glycopyrrolate (0.01 mg/kg [0.005 mg/lb], SC), oxymorphone (0.05 mg/kg [0.023 mg/lb], SC) or morphine (0.5 mg/kg [0.23 mg/lb], SC), and midazolam (0.2 mg/kg [0.09 mg/lb], SC). Anesthesia was induced with fentanyl (6 to 12 µg/kg [2.7 to 5.5 µg/lb], IV) and midazolam (0.2 mg/kg, IV) with or without etomidate (0.5 mg/kg, IV) or propofol (1.8 to 3.6 mg/kg [0.8 to 1.6 mg/lb], IV). Anesthesia was maintained with inhalation of isoflurane, infusion, or intermittent boluses of fentanyl citrate (20 to 45 µg/kg/h [9 to 20 µg/lb/min], IV) or 5 µg/kg [2.3 µg/lb], IV, respectively), and intermittent administration of atracurium besylate (0.1 to 0.25 mg/kg [0.045 to 0.11 mg/lb], IV). Positive pressure ventilation was maintained throughout the procedure except during full cardiopulmonary bypass. During this period, isoflurane was administered via a vaporizer placed in the gas line supplying the membrane oxygenator. Dexamethasone sodium phoshate (1 mg/kg [0.45 mg/lb], IV) was administered prior to cardiopulmonary bypass. Cefoxitin (22 mg/kg [10 mg/lb], IV) was administered every 90 minutes during surgery.

A 7-F triple-lumen catheter was placed percutaneously into a jugular vein to provide 2 lumens for central venous access and 1 to monitor central venous pressure (CVP). A dorsal pedal arterial catheter was placed to monitor direct arterial pressure and for arterial blood gas analysis. The electrocardiogram, arterial blood pressure, CVP, end-tidal CO₂, and esophageal and rectal temperatures were monitored continuously during surgery. Arterial and venous blood gases, activated clotting time, Hct, and concentrations of sodium, potassium, ionized calcium, and total protein were measured periodically throughout the procedure.

Dogs were positioned in dorsal recumbency for cardiopulmonary bypass and surgery. Prior to cannula for cardiopulmonary bypass, heparin sulfate (3 mg/kg [1.4 mg/lb], IV) was administered to achieve complete anticoagulation (activated clotting time > 480 sec). The left femoral artery was surgically exposed through an inguinal incision and a straight arterial cannula (8- to 12-F) placed in the artery. The heart and great vessels were exposed through a median sternotomy. Pericardium was opened on its ventral midline and secured with sutures to the sternotomy. Purse-string sutures were placed in the right atrial appendage, and a straight venous cannula was introduced into the right atrium through the right atrial appendage. In 5 dogs, an additional angled venous cannula was introduced through a pursestring directly into the cranial vena cava. Cardiopulmonary bypass was initiated by diverting blood from the right side of the heart to the cardiopulmonary bypass circuit by means of the venous cannulae.

The primary bypass circuit consisted of a roller pump, membrane oxygenator, reservoir, heat exchanger, and circulating heater/cooler water bath. The heater/cooler water bath was used to control body temperature by means of a heat exchanger built into the primary circuit. Blood in the operative field was salvaged and returned to the reservoir by 2 suction lines driven by additional roller pumps. The arterial line, vent line, and suction lines were constructed from 0.25-in tubing and connectors, whereas the venous side of the bypass circuit was constructed of 0.375-in tubing and connectors.

The bypass circuit was primed with balanced pH-adjusted (7.4) crystalloid solution. Additives to the crystalloid prime included mannitol (0.5 g/kg [0.22 g/lb]), sodium bicarbonate (15 mg/L of prime), and heparin (1,000 U/L of prime). Dogs were hemodiluted by mixing their blood volume with the circuit prime and cooled to a rectal temperature of 28 C (82.4 °F). Perfusion flows were 50 to 80 ml/kg/min [22.7 to 36.4 ml/lb/min] depending on body temperature. Phenytoin (0.05 to 0.1 mg/kg, IV) was administered periodically to increase vascular resistance and maintain mean arterial pressure above 50 mm Hg during cardiopulmonary bypass. Metabolic acidosis during cardiopulmonary bypass was corrected by administration of sodium bicarbonate (0.5 to 1 mEq/kg [0.22 to 0.45 mg/lb]) into the bypass circuit. In these same 5 dogs, a cannula was introduced through a buttressed mattress suture in the ascending aorta for administration of cardioplegia solution. The ascending aorta was cross-clamped, and complete cardiac arrest was achieved by administration of cold cardioplegia solution (15 ml/kg [6.8 ml/lb]) into the coronary circulation via the aortic cannula. Administration of cardioplegia solution was repeated every 20 minutes to maintain cardiac arrest. Cardioplegia solution consisted of balanced pH-adjusted (7.4) crystalloid solution with added potassium chloride (100 mEq/L for initial dose, 50 mEq/L all subsequent doses), sodium bicarbonate (50 mEq/L), mannitol (3 g/L), and lidocaine (100 mg/L). Heparinized blood from the bypass circuit was added to the crystalloid cardioplegia solution at a 4:1 ratio (crystalloid:blood) to make the final
Sanguineous-crystalloid cardioplegia solution. In these dogs, a left ventricular vent was introduced through a pledget-butressed mattress suture in the left ventricular apex. In the remaining 2 dogs, cardiac repair was performed on the beating heart without cross-clamping the aorta or cardioplegic arrest. In both of these dogs, ventricular fibrillation occurred during the repair and was allowed to continue until the repair was completed.

Cardiac repair—The cardiac repair was accomplished through an incision in the right ventricle that spanned the region of the defect (Fig 2). The location of the ventricular obstruction was determined by visual inspection, palpation of the right ventricular wall, or both. The fibromuscular membrane was excised, taking care to avoid injury to the papillary apparatus of the tricuspid valve. The ventriculotomy was closed by imposition of an oval-shaped polytetrafluoroethylene cardiovascular patch by use of polypropylene suture in a simple continuous pattern. The patch-graft was reinforced with pledget-butressed mattress sutures.

After cardiac incisions were closed, the heart was electrically defibrillated if necessary with direct current (20 to 50 J) by use of internal paddles. Dogs were rewarmed to 38 C [100.4 F] and gradually weaned from cardiopulmonary bypass. During this period, calcium chloride (10 mg/kg [4.5 mg/lb], IV) was administered to maintain the pH-corrected ionized calcium concentration between 1.0 and 1.2 mmol/L. Inotropic support with dobutamine (1 to 10 µg/kg/min [0.45 to 4.5 µg/lb/min], IV) or epinephrine (0.05 to 0.5 µg/kg/min, IV) was administered as necessary to support systemic blood pressure and cardiac output. Phenylephrine (0.1 to 0.2 µg/kg/min [0.04 to 0.09 µg/lb/min], IV) was also administered as needed to maintain systemic blood pressure during the weaning period. Bolus administration of lidocaine (2 to 4 mg/kg [0.9 to 1.8 mg/lb], IV) was followed by a constant rate infusion (50 to 80 µg/kg/min [22.7 to 36.4 µg/lb/min], IV), which was maintained at least until the patient recovered from anesthesia. After discontinuation of cardiopulmonary bypass, residual blood volume in the reservoir was returned via the arterial line until the CVP was between 4 and 10 mm Hg. Once the dog had been weaned from cardiopulmonary bypass and was hemodynamically stable, cannulae were removed in the reverse order that they were introduced, and protamine sulfate (0.8 to 1.2 mg/mg of heparin, IV) was administered slowly (10 to 15 minutes) to reverse the anticoagulation. Fresh whole
blood (1 unit) was administered after protamine administration was complete.

**Results**

A discrete membrane was identified on echocardiography in each dog at the base of the RVOT between 2.5 and 3.5 cm below the pulmonic valve. Right ventricular hypertrophy was present proximal to the defect in each dog and was subjectively rated as mild in 3 dogs, moderate in 2 dogs, and severe in 2 dogs. Mean maximal instantaneous systolic pressure gradient across the defect was 123 ± 89 mm Hg.

Six of 7 dogs survived surgical repair of DCRV without major complications. One dog underwent cardiac arrest during surgery before cardiopulmonary bypass was initiated. Cardiac massage was performed until cardiopulmonary bypass could be instituted. The surgery was completed, and the dog recovered. Brain death was confirmed by brain stem activity-evoked response in this dog on the day after surgery, and the dog was euthanatized. Two dogs developed pleural effusion between 3 and 4 days after surgery. Right ventricular function appeared subjectively depressed in 1 of these dogs. The effusion was responsive to treatment with enalapril (0.25 to 0.5 mg/kg [0.11 to 0.23 mg/lb], q 12 h) and furosemide (1.5 to 2.5 mg/kg [0.68 to 1.1 mg/lb], q 12 h). One dog had ventricular ectopy after surgery that was treated for 1 week with procainamide (25 mg/kg [11.4 mg/lb], q 8 h). Sinus rhythm was restored at surgery in the 2 dogs with atrial flutter or atrial fibrillation at initial examination. Atrial flutter or atrial fibrillation did not recur after surgery. Mean instantaneous pressure gradient across the defect was decreased after surgery in dogs for 1 week. Mean percentage decrease in PG after surgery was 71% (range, 40 to 94%).

Six dogs were discharged from the hospital a median of 4.3 days after surgery. Four dogs were alive and believed to be clinically normal by their owners between 2 and 56 months after surgery. One dog died unexpectedly 48 months after surgery without apparent clinical signs prior to death. One dog remained exercise intolerant for at least 6 months after surgery. Residual PG across the defect remained unchanged from the initial PG measured after surgery in dogs for which near- or intermediate-term follow-up echocardiography could be obtained. Tricuspid regurgitation decreased in severity from moderate to trivial in 1 dog and from severe to moderate in another dog after surgery. The status of tricuspid regurgitation in a third dog was not determined after surgery.

**Discussion**

In humans, DCRV is characterized by a hypertrophied muscle bundle that divides the right ventricle into a hypertrophied inflow chamber and a normal thin-walled outflow tract.9 This abnormal bundle has been theorized to represent accentuated septoparietal trabeculations or an abnormal moderator band.9 The muscle bundle is usually covered by fibrous tissue and has the appearance of a small circular orifice within the right ventricle. The defect has also been referred to as mid-right ventricular obstruction.8 In 2 reports describing DCRV in dogs,10 the obstruction was described as a thick muscular band extending from the ventricular free wall to the septum in the region of the right ventricular outflow tract. In the dogs reported here, the obstruction was a distinct muscular band covered with varying amounts of fibrous tissue, especially over the internal margin. In each instance, the defect was just distal to the tricuspid papillary apparatus and distinctly proximal (by at least 2.5 cm) to the pulmonic valve. An indentation on the outer right ventricular wall was usually present at the level of the defect. The pulmonic valve was considered normal in 6 of the 7 dogs.

Despite 2 previous reports, DCRV is not widely recognized as a distinct cardiac anomaly among dogs. The defect likely is often identified as a variation of subvalvular pulmonic stenosis (PS). Double-chambered right ventricle and PS likely share similar natural histories and pathophysiologic consequences. Nevertheless, a distinction between DCRV and PS may be important for several reasons, including apparent differences in breed prevalence and treatment implications for the 2 defects. Pulmonic stenosis is overrepresented in several terrier breeds, English Bulldogs, and Chihuahuas.10 In contrast, 5 of 7 dogs with DCRV in our report were Boxers or Golden Retrievers, neither of which were identified as having substantially increased risk for PS.10 This supports the conclusion that PS and DCRV are likely distinct cardiac defects with different patterns of heritability.

Several options have been used successfully for treatment of PS, including balloon valvuloplasty, surgical dilation valvuloplasty, and transannular patch-graft valvuloplasty performed under inflow occlusion. Balloon valvuloplasty is preferred for palliation of PS, because it is less invasive and has a reasonably high success rate.11 Balloon valvuloplasty was attempted without success in 4 dogs of this report, either because a balloon could not be passed across the obstruction or because balloon dilation failed to appreciably relieve the obstruction. Although inflow occlusion has been successfully used for surgical correction of PS,12 the nature and location of the DCRV defect was not believed to be amendable to correction under inflow occlusion alone. Surgical correction of DCRV was undertaken during cardiopulmonary bypass to allow adequate time to discern and excise the obstruction and to reconstruct the ventricular wall spanning the defect. Thus, DCRV and PS appear to have important differences and implications with regard to options for treatment.

Diagnosis of DCRV in humans is based primarily on 2-dimensional and Doppler echocardiographic findings.7 Less common means of confirming a diagnosis of DCRV include selective angiography,6 magnetic resonance imaging,12 and scintigraphy.12 Two-dimensional and Doppler echocardiography were believed to be reliable and sensitive methods of diagnosis for DCRV in the dogs of this report.

Defects associated with DCRV in humans include ventricular septal defect, atrial septal defect, double-outlet right ventricle, pulmonary valve stenosis, and discrete subvalvular aortic stenosis.7 Six of 7 dogs in
this report had at least 1 concurrent cardiac defect. Four dogs had moderate or greater tricuspid regurgitation, which was believed to be important because of its probable exacerbating effect on congestive heart failure. Three of these dogs had restrictive leaflet motion suggestive of congenital tricuspid valve dysplasia. Follow-up echocardiographic evaluation was available in 2 dogs with tricuspid regurgitation; in both dogs, tricuspid regurgitation decreased substantially within a few months of surgery. This finding emphasizes the important effect that increased right ventricular pressure has on tricuspid regurgitation. Further, it was believed to have important implications regarding early surgical intervention for dogs with DCRV and tricuspid regurgitation, even when the tricuspid valve is dysplastic.

In humans, this anomaly is typically progressive, resulting in increasing pressure gradients and right ventricular functional impairment if it is not treated in a timely fashion. The defect potentially causes life-threatening arrhythmias secondary to myocardial ischemia, subsequent endocardial fibrosis, and increased risk for sudden cardiac death. Early surgical correction of a substantial obstruction routinely results in an excellent clinical outcome. Five of the 7 dogs of this report had clinical signs of syncope, congestive heart failure, or both at an early age. Surgical correction of DCRV resulted in an excellent clinical outcome in all but 1 dog that survived surgery. One dog did not survive surgery, and 1 dog remained exercise intolerant after surgery. Both of these dogs were in refractory congestive heart failure at the time of surgery. One dog died unexpectedly 48 months after surgical correction without warning or apparent clinical signs. This dog had documented ventricular tachycardia before and after surgery. These findings suggest that surgical intervention can be expected to have a favorable clinical outcome in dogs with DCRV, particularly if it is undertaken before the onset of congestive heart failure. As with other obstructive-type cardiac defects, some retained risk for sudden cardiac death should be expected.

In conclusion, DCRV is a congenital heart defect of dogs that may emerge in importance as awareness of the condition as a distinct anomaly increases. Although DCRV shares pathophysiologic features with pulmonic stenosis, diagnosis of DCRV likely has important implications with regard to breed prevalence and treatment options. Surgical correction of DCRV improved exercise tolerance in most dogs of our report. Congestive heart failure before surgery may be associated with a less than optimal outcome. A survival benefit associated with surgical correction of DCRV cannot be determined from results of this study.

References