J Vet Intern Med 2005;19:262–267

Double-Outlet Right Ventricle in an Angus Calf

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Double-outlet right ventricle (DORV) is a conotruncal malformation where both great arteries arise from the right ventricle. Anatomic variations of DORV are classified according to the position of the great arteries in relation to each other, the relationship between a ventricular septal defect (VSD) and the great arteries, and the presence and degree of pulmonary stenosis.[1] The prevalence of congenital cardiac defects in bovine fetuses has been reported at approximately 0.7 %, with VSDs representing the most common congenital cardiac defect.[2] DORV has been described in veterinary literature in few cats and dogs,[3–6] a foal,[7] and 2 calves[8,9] with variable clinical and pathologic documentation. In this report, we describe the angiographic, echocardiographic, and postmortem examination findings in a calf with a DORV with concurrent pulmonary stenosis, subaortic VSD, patent ductus arteriosus (PDA), aberrant left subclavian artery, and a tracheal malformation.

A 1-day-old female Angus calf was presented to the University of Illinois Veterinary Teaching Hospital for recumbency and tachypnea. The calf was delivered without complications. There was no known exposure to chemical or viral agents during gestation. The calf was weak and unable to stand on presentation and a weak suckle response was present. A continuous murmur was noted over the left heart base and a pansystolic crescendo-decrescendo murmur was noted cranially on the right hemithorax. The results of auscultation of the lungs were normal; however, modest tachypnea and dyspnea were noted. Temperature was 98.3°F (reference range, 100–102°F) and the peripheral pulse rate matched the heart rate at 64 pulses/min (reference range, 90–110 pulses/min). Both jugular veins were distended and a prominent jugular pulse was noted. The calf's blood glucose was 31 mg/dL (reference range, 80–100 mg/dL), and PCV was 32% (reference range, 28–40%) with a total protein of 4.0 g/dL (reference range, 5.7–7.1 g/dL). A serum chemistry profile revealed sodium of 143 mEq/L (reference range, 132–138 mEq/L), potassium of 4.1 (reference range, 4.9–6.1 mEq/L), chloride of 108 mEq/L (reference range, 96–109 mEq/L), ionized calcium of 4.2 mg/dL (reference range, 4.2–5.3 mg/dL), and pH of 7.46. Ten milliliters of 50% dextrose was administered via the left jugular vein and lactated Ringer's solution with 5% dextrose was started via the left ear vein at a maintenance rate (60 mL/h). The calf nursed 1 L of dam colostrum and an additional 3 L were given via an oroesophageal feeder tube. Hypoglycemia and hypothermia resolved within several hours but the calf remained moribund.

Echocardiographic findings included a severely dilated and hypertrophied right ventricle with moderate right atrial enlargement and a large VSD inferior to the crista supraventricularis (Fig 1). Color-flow and spectral Doppler demonstrated low-velocity, bidirectional shunting through the VSD. The shunting was left to right during systole and right to left during diastole, with velocity less than 0.75 m/s in each direction, suggesting equalization of left and right ventricular pressures (Fig 2). A slightly oblique right parasternal short-axis view revealed 2 great vessels leaving the right ventricle, with the larger vessel (assumed to be an overriding aorta) measuring 2.2 times the diameter of the smaller vessel (Figs 3, 4). Turbulent flow of high velocity (4.2 m/s) was noted in the smaller vessel and was thought to represent a hypoplastic pulmonary artery with a suspected valvular stenosis. Right parasternal long-axis view revealed a distorted interventricular septum in the area of the VSD (Fig 5). Other notable findings included modest tricuspid insufficiency, a patent foramen ovale, and an enlarged left coronary artery leaving its respective sinus of Valsalva. The cause of the continuous murmur could not be identified.

Anesthesia was induced with and maintained by using isoflurane mixed with 100% oxygen. Angiographic 7F pigtail catheters[a] were placed in the right jugular vein and carotid artery by means of a cutdown. When advanced under fluoroscopic guidance, both catheters entered the right ventricle. A right ventricular angiogram[b] was performed and demonstrated dilation and hypertrophy of the right ventricle, with blood flow directed out a hypoplastic pulmonary artery and a dilated cranial and dextropositioned ascending aorta (Fig 6). Mild tricuspid valve insufficiency was present. Injection into the aortic root showed modest flow through a PDA, dilation of both sinuses of Valsalva and the initial part of both coronary arteries, and an aberrant left subclavian artery. The calf was euthanized and a postmortem examination was performed.

The heart was enlarged and globoid in appearance. The ascending aorta lay to the right and cranial of the hypoplastic pulmonary trunk (Fig 7). Aberrant origin of the left subclavian artery was evident along with a probe-PDA. A window was made by removing a segment of the right ventricular wall, allowing visualization of the abnormal aortic origin from the right ventricle (Figs 8, 9). The only outlet for the left ventricle was a large VSD, which was subaortic and inferior to the crista supraventricularis. The right ventricle was hypertrophied and the conus of the pulmonary artery was narrowed and underdeveloped, consistent with a severe form of subpulmonic stenosis (Figs 8, 9). Inspection of the pulmonary trunk revealed pulmonic valve leaflets that were thickened, fused, and stenotic (Fig 9). No continuity was found between the aortic root and mitral valve. Other notable findings were an incomplete fusion of

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Submitted July 15, 2004; Revised October 12, 2004; Accepted November 12, 2004.

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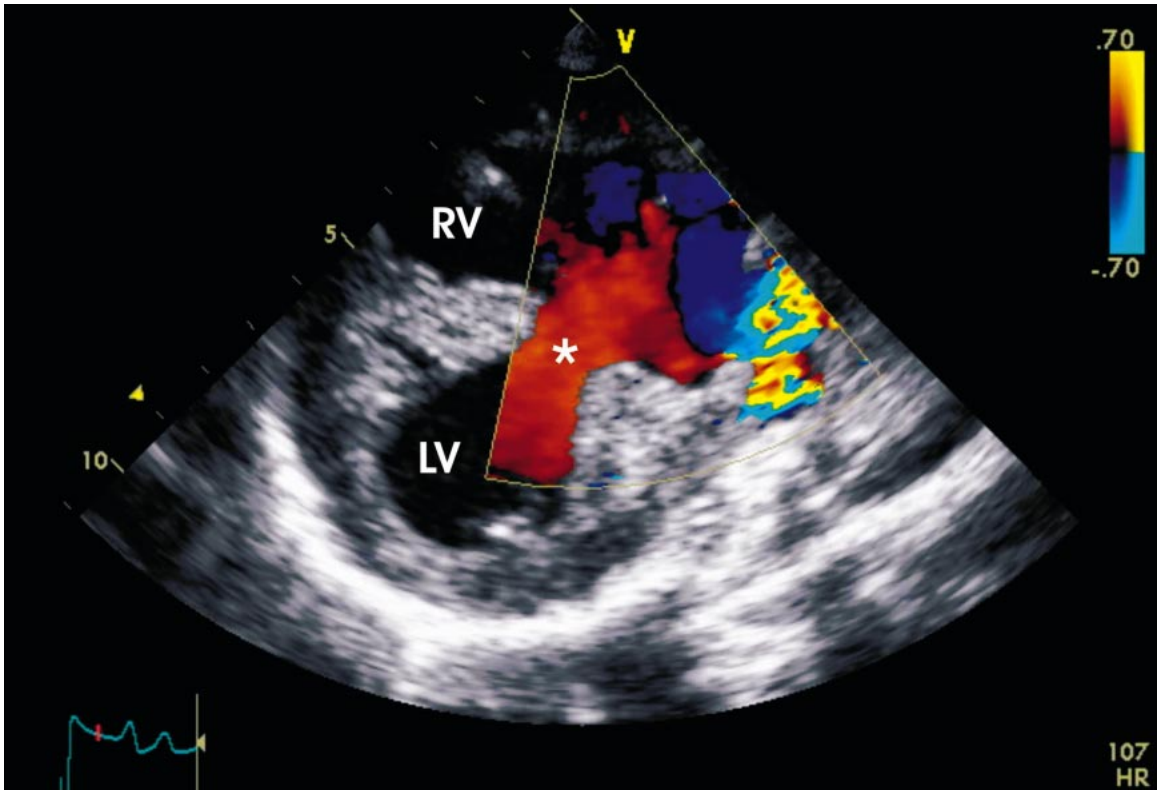

Fig 1. Right parasternal short-axis view displaying the presence of a large ventricular septal defect (VSD) and systolic shunting of blood from the left to right ventricle. A mosaic color Doppler display originates just below the origin of the hypoplastic pulmonary artery, consistent with subpulmonic stenosis. RV, right ventricle; LV, left ventricle; *, VSD.

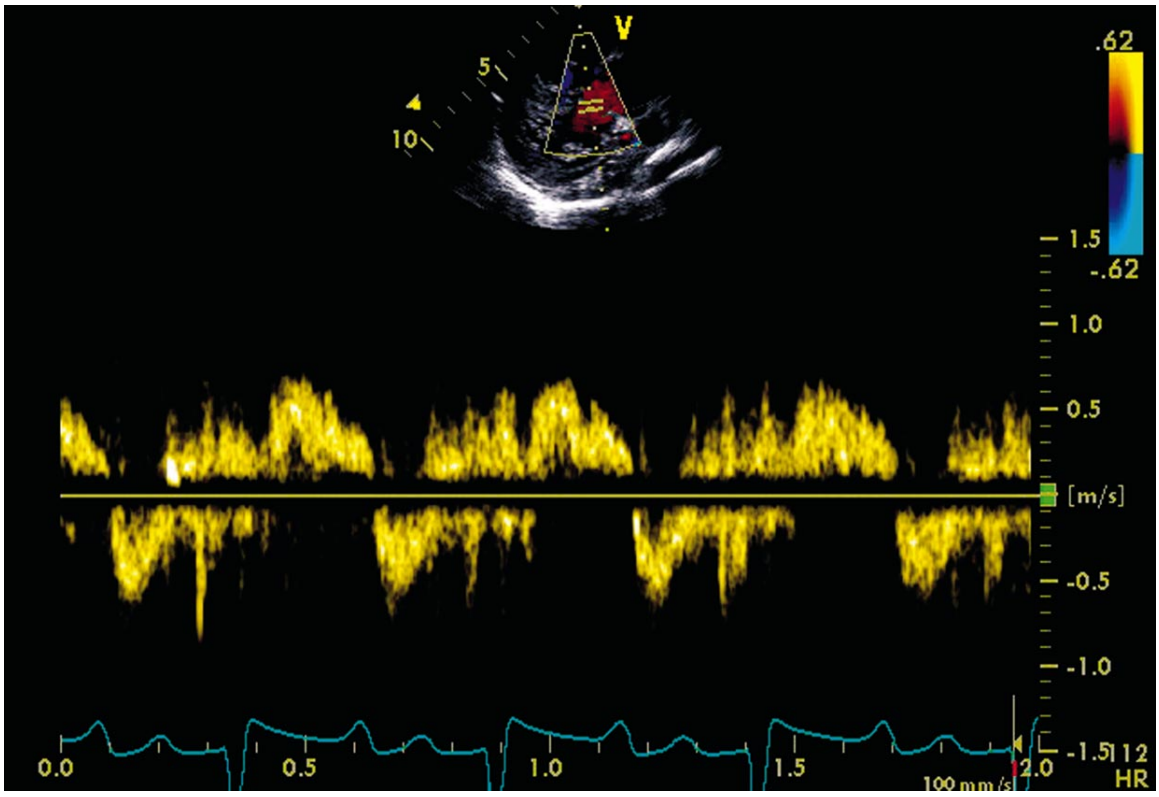

Fig 2. Continuous Doppler study across the ventricular septal defect demonstrating bidirectional low-velocity shunting. Direction of flow is left to right during systole and right to left during diastole.

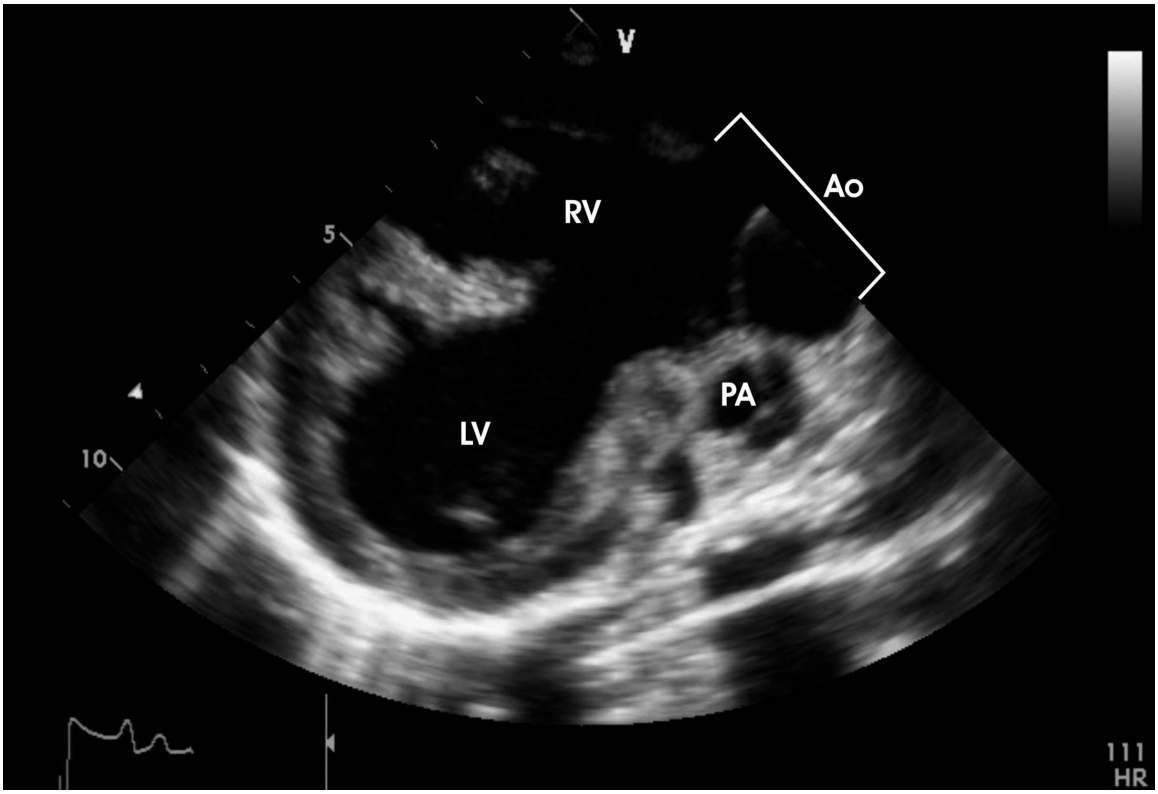

Fig 3. Oblique right parasternal short-axis view showing the aorta (Ao) and hypoplastic pulmonary artery (PA) arising from the right ventricle (RV). Note the large discrepancy in diameter of the aorta and pulmonary artery. A large ventricular septal defect is seen connecting the left ventricle (LV) with the right ventricle.

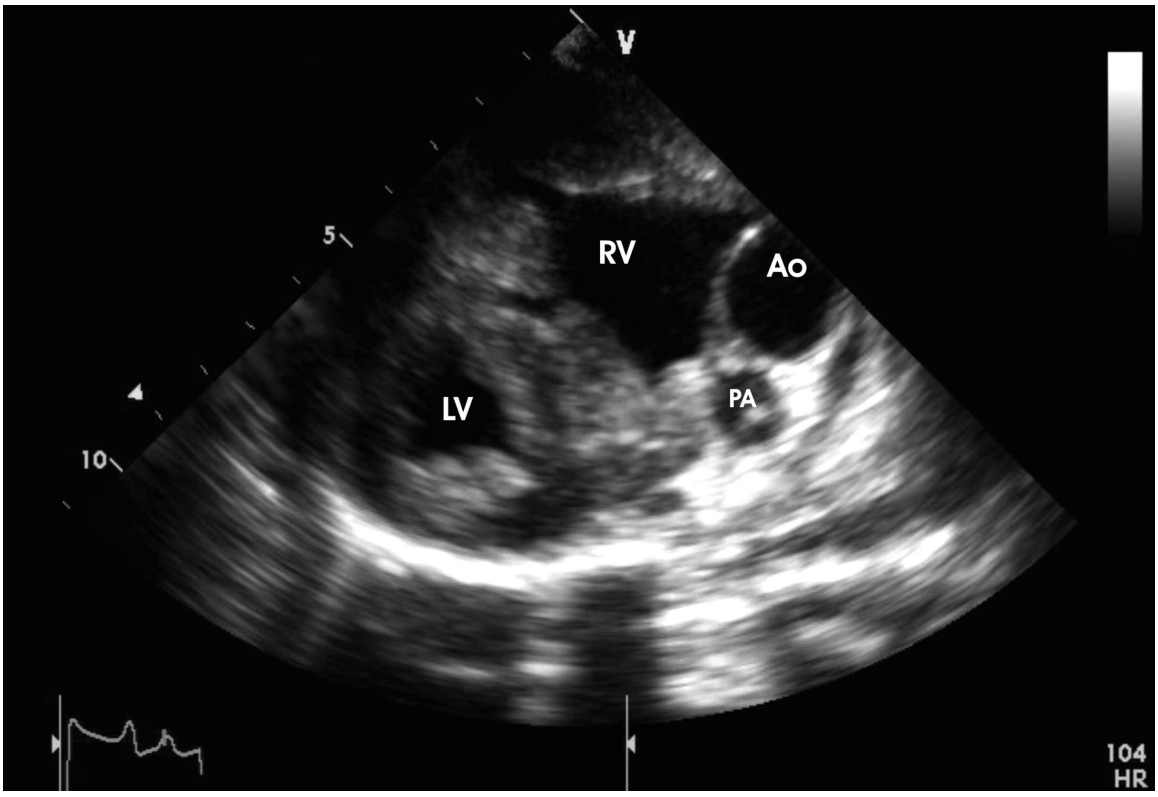

Fig 4. Oblique right parasternal short-axis view confirming the origin of 2 great vessels, the aorta (Ao) and pulmonary artery (PA), from a hypertrophied and dilated right ventricle (RV). LV, left ventricle.

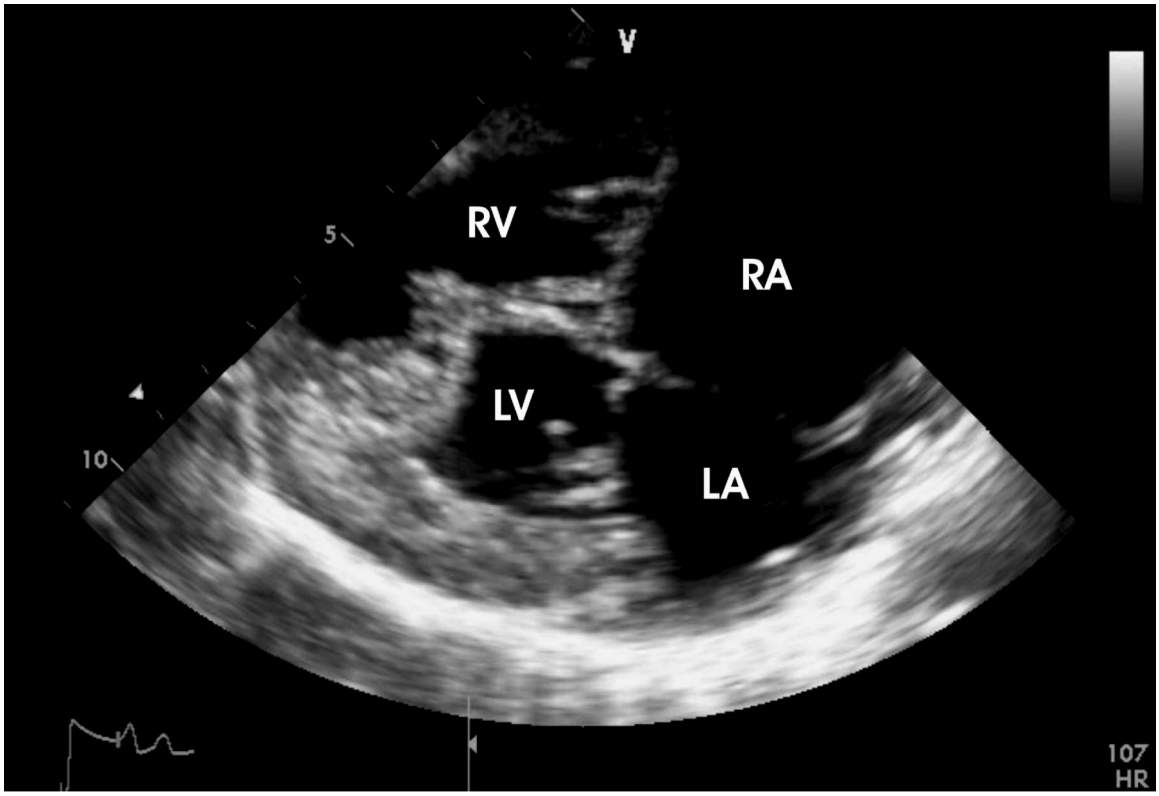

Fig 5. Right parasternal long-axis view displaying a dilated right atrium (RA) and dilated and hypertrophied right ventricle (RV) with a distorted interventricular septum and papillary muscle due to a ventricular septal defect. LV, left ventricle; LA, left atrium.

the foramen ovale flap and a hypoplastic trachea with fusion of its dorsal rings giving a teardrop-shaped appearance on cross-section (Fig 10).

The incidence of DORV in humans is 1–1.5% of patients with congenital heart disease.[10] There are few reports of DORV in domestic animals and this defect is assumed to be rare.[3–9] The prognosis for DORV in animals appears to be poor but one successful surgical repair of DORV has been reported in a cat.[4]

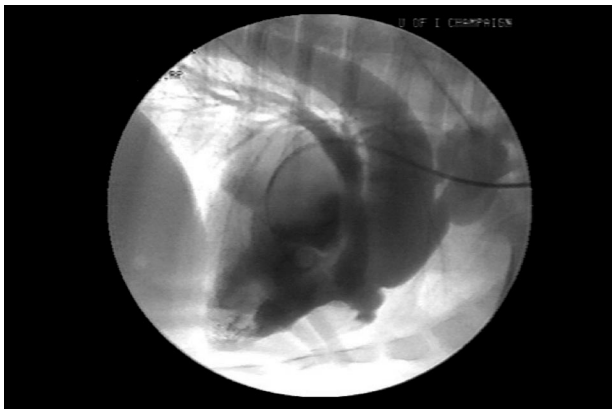

Fig 6. Right ventricular angiographic injection with opacification of a hypoplastic pulmonary trunk and a dilated aortic root. Modest tricuspid insufficiency and moderate right atrial enlargement are evident. Branching of the aorta is abnormal and a suspected aberrant left subclavian is present. Moderate right ventricular hypertrophy and dilation also are noted.

The antemortem distinction between DORV from other conotruncal abnormalities such as the tetralogy of Fallot and complete transposition of the great vessels is made difficult because of the common embryologic origin and close relationship of these defects.[11] A diagnosis of DORV is made when both great vessels arise from a distinct conus originating from the morphologic right ventricle and from which no fibrous continuity with the atrioventricular valves can be demonstrated.[1] Typically, cases of DORV are categorized according to the location of the VSD and presence or absence of pulmonic stenosis. DORV with a subaortic VSD, pulmonary stenosis, and greater than 50% aortic override resembles, both morphologically and physiologically, tetralogy of Fallot.[1] DORV with a subpulmonic VSD and no pulmonary stenosis (Taussig-Bing anomaly) resembles complete transposition of the great arteries.

A VSD provides the left ventricle its only exit in animals and humans with DORV.[1] The location of the VSD in humans is usually subaortic or subpulmonic, but less commonly the defect is committed to both great arteries (aorta and pulmonary trunk) or to neither great artery.[1] Pulmonary stenosis is present in 40–70% of humans with a subaortic VSD and is manifest as either an underdeveloped subpulmonary conus, a stenotic and often bicuspid pulmonary valve, or both. The clinical manifestations of a DORV with subaortic VSD and pulmonary stenosis closely resemble tetralogy of Fallot, with signs predominantly related to systemic hypoxemia.[1] Palliative treatment of tetralogy of Fallot or DORV includes the surgical creation of a left-to-right arterial shunt from the left subclavian to pulmonary artery

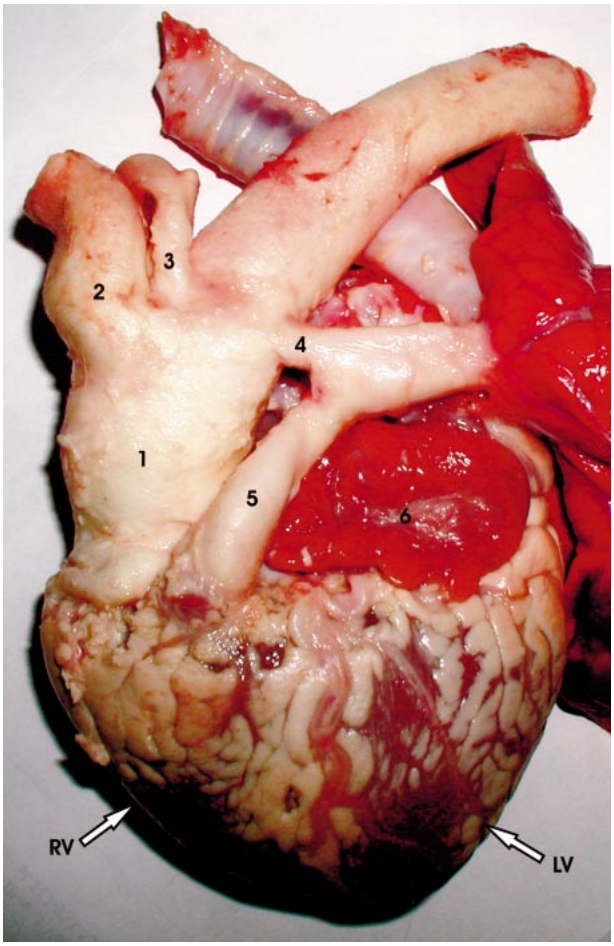

Fig 7. Gross specimen of a calf heart with moderate right ventricular enlargement, craniad and rightward positioning of an enlarged aortic root, hypoplastic pulmonary trunk, patent ductus arteriosus, and aberrant left subclavian artery. 1, aortic root; 2, brachyocephalic trunk; 3, aberrant left subclavian artery; 4, patent ductus arteriosus; 5, pulmonary trunk; 6, left auricular appendage; RV, right ventricle; LV, left ventricle.

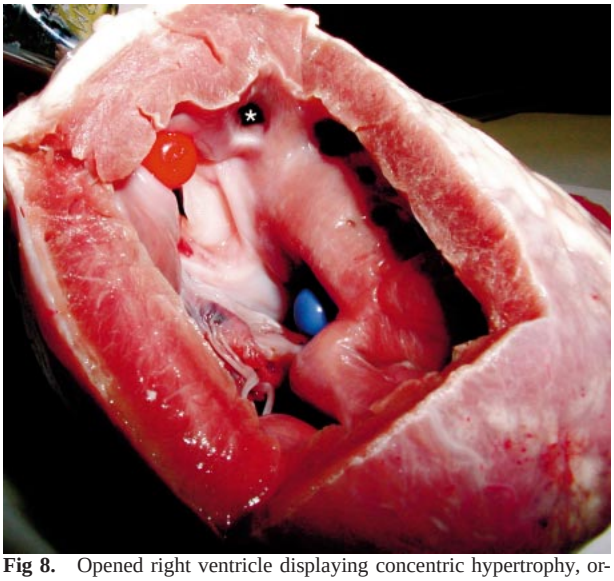

Fig 8. Opened right ventricle displaying concentric hypertrophy, origins of the aorta (red marker) and pulmonary artery (*), along with a large subaortic ventricular septal defect (blue marker). Note the severely underdeveloped and stenotic pulmonic conus.

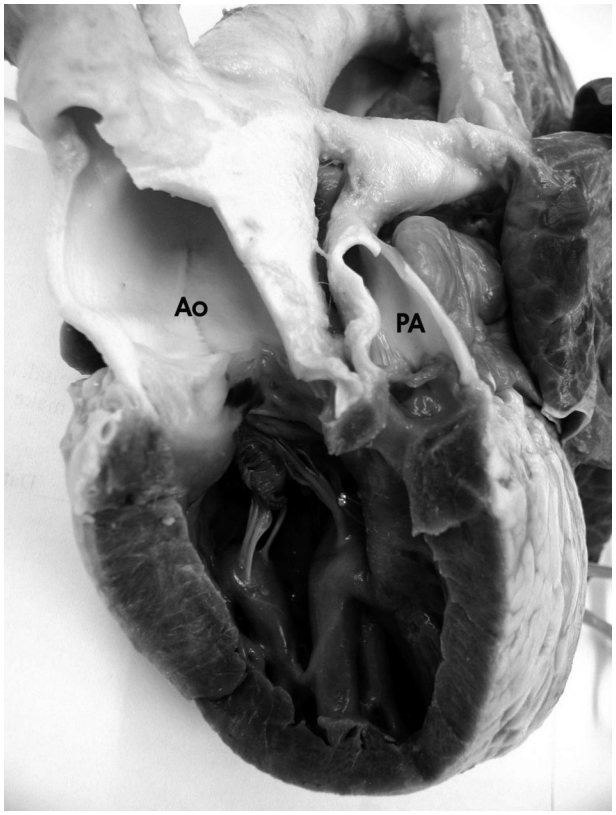

Fig 9. Opened right ventricle displaying the origin of both the aorta and pulmonic artery from the right ventricle. Note how the position of the aorta precludes any continuity of the aortic root with the mitral valve apparatus (not visible). The aortic diameter is about 2.3 times the pulmonic diameter. Pulmonic stenosis is present at the level of pulmonic valve, with leaflets that are thickened and fused and at the level of the pulmonic conus, which is severely narrowed. Moderate right ventricular hypertrophy is present. An aberrant left subclavian artery and patent ductus arteriosus also are noted. Ao, aorta; PA, pulmonary artery.

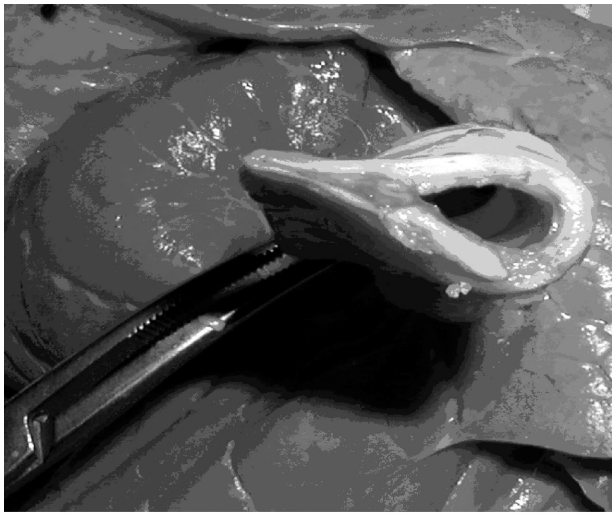

Fig 10. Hypoplastic trachea with fusion of its dorsal border giving a teardrop-shaped appearance in a calf with a double-outlet right ventricle.

(Blalock-Taussig shunt). The PDA found in this calf may have acted as a similar type of shunt and presumably improved the amount of pulmonary blood flow.

The authors had some difficulty classifying the VSD as subaortic or noncommitted and the exact nature of the defect was not certain until careful inspection of the heart at postmortem examination. A particularly interesting finding was the malformed teardrop-shaped hypoplastic trachea, which the authors have not previously seen. In humans, tracheal stenosis and tracheal agenesis with DORV has been reported; however, the exact description of the malformation found in this calf could not be found.[12,13] The presence of this malformation may have contributed to the calf's dyspnea. In a species in which diagnostics are often limited because of financial constraints, the correlation of clinical and pathologic features of DORV, as presented in this report, should aid the future recognition of this uncommon disorder.

Footnotes

[a] Catheter, Terumo Medical, Somerset, NJ
[b] Hypaque, Amersham Health, Princeton, NJ

References

1. Perloff JK. Double outlet ventricle. In: Perloff JK, ed. Clinical Recognition of Congenital Heart Disease. Philadelphia, PA: WB Saunders; 2003:383–402.

2. Kemler AG, Martin JE. Incidence of congenital cardiac defects in bovine fetuses. Am J Vet Res 1972;33:249–251.

3. Abduch MC, Tonini PL, Oliveira Domingos BL, et al. Double-outlet right ventricle associated with discordant atrioventricular connection and dextrocardia in a cat. J Small Anim Pract 2003;44:374–377.

4. Northway RB. Use of an aortic homograft for surgical correction of a double-outlet right ventricle in a kitten. Vet Med Small Anim Clin 1979;74:191–192.

5. Jeraj K, Ogburn PN, Jessen CA, et al. Double outlet right ventricle in a cat. J Am Vet Med Assoc 1978;173:1356–1360.

6. Van der Linde-Sipman JS, van den Ingh TS, Koeman JP. Congenital heart abnormalities in the cat: A description of sixteen cases. Zentralbl Veterinarmed 1973;20A:419–425.

7. Chaffin MK, Miller MW, Morris EL. Double outlet right ventricle and other associated congenital cardiac anomalies in an American Miniature Horse foal. Equine Vet J 1992;24:402–406.

8. Wilson RB, Cave JS, Horn JB, Kasselberg AG. Double outlet right ventricle in a calf. Can J Comp Med 1985;49:115–116.

9. Zulauf M, Tschudi P, Meylan M. [Double outlet right ventricle (DORV) in a 15 month old heifer.] Schweiz Arch Tierheilkd 2001; 143:149–154.

10. Silka MJ. Double outlet right ventricle. In: Garson A, Bricker JT, Fisher DJ, Neish SR, eds. The Science of Pediatric Cardiology. Philadelphia, PA: Williams & Wilkins; 1998:1505–1523.

11. Lomonico MP, Bostrom MP, Moore GW, Hutchins GM. Arrested rotation of the outflow tract may explain tetralogy of Fallot and transposition of the great arteries. Pediatr Pathol 1988;8:267–281.

12. Oshima Y, Yamaguchi M, Ohashi H, et al. [Pulmonary artery sling with tracheal stenosis–Primary repair in infancy.] Jpn J Thorac Cardiovasc Surg 1998;46:347–353.

13. Oppido G, Napoleone CP, Loforte A, et al. Complex double-outlet right ventricle repair in a neonate with complete tracheal agenesis. J Thorac Cardiovasc Surg 2004;127:283–285.