Double-inlet Univentricular Heart of Left Ventricular Type in a Calf: Detection by Two-dimensional and Doppler Echocardiography

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The term "double-inlet univentricular heart" used in this report includes hearts in which the 2 atria connect to only 1 chamber (main chamber; MC), with or without a second chamber (rudimentary chamber; RC) receiving no atrioventricular inlet [1]. Traditionally, this malformation has been termed "cor triloculare, biatriatum". Based on the morphology of the MC and RC chambers, this anomaly can be divided into 3 types: left ventricular (LV) type with a RC of right ventricular (RV) type, RV type with a RC of LV type, and indeterminate type without a RC [1]. This malformation has rarely been reported in bovine [9, 10] and equine [11] species. Two-dimensional echocardiography (2D echo) is known to be of great value for the noninvasive diagnosis of this anomaly in human patients [5, 7, 8], but there have been no reports of complete antemortem echo diagnosis in domestic animals with this condition. We report here a case of double-inlet univentricular heart of the LV type detected by 2D and Doppler echo and confirmed by necropsy.

A 9-day-old male Holstein calf weighing 45 kg was referred for evaluation of a cardiac murmur. The calf was outwardly normal. The heart rate was 115 beats/min and a grade 5/6 holosystolic murmur and thrill, with a maximum intensity at the level of the costochondral junction in the 3rd intercostal space was audible on auscultation. On blood examination, the packed cell volume (%), red blood cell count ($10^6/\mu\text{l}$) and hemoglobin concentration (g/dl) were 22, 606 and 7.1, respectively. Lateral thoracic radiography showed mild cardiomegaly with nearly normal lung vascularity. Catheterization under light sedation with xylazine revealed the following pressure (systole/diastole, mm Hg) and PO_2 (mm Hg) values: right atrium, 9/-2, 29; large ventricular chamber, 164/0, 37; aorta, 163/100, 45.

2D echo (Hitachi EUB-40, Hitachi Medical

Corp., Tokyo) examination was performed using our systematic 2D echo approach for the diagnosis of congenital heart disease in cattle [2]. A modified apical long-axis view of the heart obtained from the left 4th intercostal location demonstrated drainage of the posterior vena cava to the normally positioned right-sided atrium, indicating that this atrium was the morphologic right atrium; this was confirmed by subsequent contrast echo following injection of contrast agent into the saphenous vein. Long-axis 4-chamber views of the heart from the left and right 4th intercostal space also revealed pulmonary venous drainage to the normally positioned left-sided atrium, indicating that this atrium was the morphologic left atrium. Thus, atrial-visceral situs was diagnosed as solitus. From the same views as those above, 2 distinct atrioventricular valves were seen to open into a large single chamber (*i.e.*, the MC) without any apparent interventricular septum (Fig. 1), indicating a double-inlet atrioventricular connection. Serial short-axis views of the LV from apex to base obtained from the right 4th intercostal scan clearly showed the anatomical three-dimensional structure of this heart (Fig. 2). Up to the level of the papillary muscle, only 1 ventricle (the MC) was found (Fig. 2A). Further dorsal tilting of the transducer revealed a septum (trabecular septum) and a small chamber (*i.e.*, the RC) lying cranial to the MC (Fig. 2B). Further dorsal tilting showed 2 atrioventricular valve orifices within the MC and a septal defect (outlet foramen) approximately 1 cm wide (Fig. 2C). By further dorsal tilting of the transducer, one great artery could be seen arising from the MC, while another great artery arose from the RC (Fig. 2D). From these short- and long-axis views, the MC was presumed to be a morphologic LV simply on the basis of its smoother trabecular pattern. Identification of each great artery was made on the basis of its morphologic features together with the spatial relationships of both great arteries using a series of left 3rd intercostal short-axis

Fig. 1. (A) A long-axis four-chamber view of the heart demonstrating two separate atrioventricular valves opening into one ventricle (the main chamber: MC). (B) With the transducer angled somewhat more craniad, a great artery (aorta: Ao) is seen originating from the MC. L=left atrioventricular valve; LA=left atrium; R=right atrioventricular valve; RA=right atrium.

views (Fig. 3). The presumed aorta arose from the MC, while the presumed pulmonary artery (PA) arose from the RC, and a normal great arterial relationship was noted, indicating a concordant ventriculo-arterial connection. From these 2D echo data, the claf was diagnosed as having a double-inlet univentricular heart of the LV type with a RC of the RV type.

Color Doppler echo (Toshiba SSH-65A, Toshiba Medical Corp., Tokyo) revealed a stream of blood (shunt flow) crossing the septal defect from the MC to the RC which occurred twice during the cardiac cycle, once in systole and again in diastole (Figs. 4A, B), which was confirmed by subsequent pulsed Doppler echo with the sample volume placed in the RC adjacent to the defect. Additional color Doppler echo findings included 2 separate atrioventricular diastolic inflows characterized by normal speed and nonturbulence directed toward the MC apex (Fig. 4C), verifying the absence of atresia or stenosis of both atrioventricular valves, which is not infrequently seen in this type of cardiac anomaly [1], and mild degrees of aortic and

Fig. 2. Serial short-axis views of the ventricle from apex to base. (A) Papillary muscle level. (B) Chordal level. (C) Atrioventricular valve level. (D) Ventricular outflow tract level. (E) Semilunar valve level. For explanations of findings, see text. AV=aortic valve; C=chordae tendineae; D=septal defect; P=papillary muscle; PV=pulmonary valve; RC=rudimentary chamber; S=trabecular septum; Other abbreviations as in Fig. 1.

Fig. 3. Serial short-axis views of the great arteries from the atrioventricular valve (panel A) to the supra-semilunar valvular level (panel D). (A) Two atrioventricular valves are seen entering a large single chamber (main chamber:MC). (B) Further dorsal tilting of the transducer reveals a ventricular outflow tract (VOT) beneath an arterial valve committed to the MC. A cranially located, small outlet chamber (rudimentary chamber: RC) is seen in this slice, and there is a septal defect (D) between the two outlet regions. (C) With the transducer tilted more dorsally, two arterial valves of the great arteries are seen. Note that the right caudal artery (aorta: Ao) bears a coronary artery (CA). (D) Further dorsal tilting demonstrates separation of the right caudal artery into two circular echoes and the left cranial great artery (main pulmonary artery: PA) extending caudad and bifurcating into two branches. This suggests that the right caudal artery is the aorta, the left cranial artery being the pulmonary artery, and that the great arteries have a normal relationship. AA=aortic arch; BA=brachiocephalic artery; LPA=left pulmonary artery; RPA=right pulmonary artery; Other abbreviations as in previous figures.

pulmonary regurgitant flow.

On reexamination at 90 days of age, except for a slightly increased packed cell volume (32%) and disappearance of the pulmonary regurgitation, there were no remarkable changes in clinical, hematological or 2D and Doppler echo findings. This suggested that the calf still had a relatively favorable hemodynamic status, *i.e.*, favorable blood mixing of the two atrial streams within the MC and adequate pulmonary blood flow shunted from the MC through the septal defect. The calf was subsequently euthanatized and necropsied, and the antemortem diagnosis was proved to have been totally correct. Atrial situs solitus with normal venous drainage was

present. The atrial septum showed a small (4 mm in diameter) patent foramen ovale. The right and left atrioventricular orifices, with a tricuspid and mitral morphology, respectively, opened into a large chamber (the MC), which had an LV-type morphology with a smooth trabecular pattern (Fig. 5A). A small separate outlet chamber (the RC) with a coarse trabecular pattern and an absence of atrial communication was found craniodorsal to the MC (Fig. 5B). A interventricular septum (trabecular septum) separating the 2 chambers showed a small (0.9×0.6 cm) opening, *i.e.*, an interventricular communication. The aorta arose from the MC, being caudal and to the right of the PA, which arose from the

Fig. 4. Color Doppler echocardiograms. In the velocity/variance mode, color flow velocities toward and away from the transducer are depicted as red and blue, respectively. The magnitude of the velocity is represented by the brightness of the color and variance (turbulence) by the addition of green to the primary red or blue color. Highly turbulent and/or high-velocity flows result in a mixture of color (mosaic pattern). (A) A short-axis view of the heart (similar view to that in Fig. 2C)(systolic frame), showing a mosaic pattern of color reflecting high velocities and/or turbulence crossing the septal defect from the main chamber (MC) to the rudimentary chamber (RC), joining a blue-colored area representing the remainder of the RC and proximal main pulmonary artery. (B) Same view as in panel A (diastolic frame) showing a red color crossing the septal defect from the MC to the RC. Note also a mild pulmonary regurgitant flow (PR) visualized in the RC proximal to the pulmonary valve, appearing as a red color. (C) A long-axis four-chamber view of the heart demonstrating 2 separate atrioventricular diastolic inflows, appearing homogeneously red, indicating rather low velocities. Abbreviations same as in previous figures.

RC. A ventriculo-arterial connection as concordant as that in the present case is less common in human cases of double-inlet univentricular heart of the LV type [1]. The ascending aorta was larger than the main PA (4.4 cm versus 2.6 cm in external diameter).

2D echo is a very useful noninvasive method for making accurate anatomical diagnoses of various types of congenital heart defect in cattle, especially in calves [3]. The present report also demonstrates the usefulness of this method in the diagnosis of complex cardiac anomalies such as that in the present case, especially when the aforementioned systematic 2D echo diagnostic approach is carried out [2]. It has been pointed out previously that there might be considerable difficulty in confidently determining the ventricular morphology of the MC as well as that of the RC by 2D echo alone [5, 7]. In the present case, however, we were able to correctly diagnose the MC as LV type because of its relatively unequivocal trabecular appearance. In this regard, the position of the RC relative to the MC in humans is considered to be an additional useful

feature for defining the type of the MC [7]. For instance, in the case of univentricular hearts of the LV type, the RCs are usually positioned anterosuperiorly (craniodorsally) and either to the right or left of the MCs, as was seen in our present bovine case, while in the case of univentricular hearts of the RV type, the RCs are posteroinferior and to the right, left or directly posterior [7]. Color Doppler echo with conventional pulsed and continuous-wave Doppler capabilities is a newly developed item of ultrasound equipment which provides realtime blood-flow imaging superimposed on 2D echo imaging, permitting us to define and localize normal and abnormal flows within cases of the pathologic anatomy [4, 6]. In the present case, using this new technique, we were able to easily and rapidly detect the shunt flow from the MC to the RC and also determine its exact timing using conventional pulsed Doppler echo. In conclusion, the present study has demonstrated that complex cardiac anatomy detected with 2D echo is best understood when complemented by the hemodynamic information obtained using Doppler echo.

Fig. 5. Interior of the heart specimen (A) as viewed from the main ventricular chamber, and (B) as viewed from the rudimentary outlet chamber. LAO=left atrioventricular orifice; RAO=right atrioventricular orifice; Other abbreviations as in previous figures.

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要 約

断層およびドプラ心エコー図法により診断し得た左心室型単心室のウシ1例(短報): 萩尾光美・村上隆之¹⁾・大塚宏光(宮崎大学農学部家畜外科学教室, ¹⁾家畜解剖学教室)——左心室型単心室の症例(ホルスタイン種, 雌, 9日齢)に系統的な断層心エコー図診断を実施し, 生前に確定診断を得ることができた。カラードプラ法では, 主心室腔(左室)から心室間交通路を経て痕跡的心室腔(右室)へ至る収縮期および拡張期の一方向性・2相性の短絡血流が観察された。断層およびドプラ心エコー図法は本奇形のような複雑心奇形の診断・血行動態の分析に有用であると考えられた。