Three Cases of Bovine Extreme Tetralogy of Fallot

Tetsuya NAKADE, Yoshiko UCHIDA, and Kanjurou OTOMO

Department of Veterinary Surgery II, School of Veterinary Medicine, Rakuno Gakuen University, 582 Bunkyo-dai-Midorimachi, Ebetsu, Hokkaido 069, Japan

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ABSTRACT. Three female Holstein calves having cyanosis of the visible mucous membrane, exercise intolerance and systolic murmur were examined. Extreme Tetralogy of Fallot with ventricular septal defect was confirmed at autopsy. All of the intrapericardial pulmonary arteries were completely absent in two cases. In the other case, the atretic pulmonary trunk, with a bicuspid valve, rose from a very small chamber which was located on the blind-ending muscular infundibulum. There was no connection between the atretic pulmonary trunk and the right ventricle.—**KEY WORDS:** bovine, extreme tetralogy of Fallot, pulmonary atresia.

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Tetralogy of Fallot (TOF) in bovine and equine has been reported [3, 5–9, 12, 14, 15, 17], but there have only been a few case reports about extreme TOF in bovine and equine [10, 13, 16, 18, 19]. This paper describes three cases of bovine severe TOF which were confirmed post-mortem. We were able to make definitive diagnosis in two cases through the use of various kinds of antemortem examinations, but were not able to make a proper diagnosis in one case.

Case 1 (No. 1590)

An 8-day-old female Holstein calf was admitted to the Veterinary Medical Teaching Hospital, Rakuno Gakuen University, because of a decrease in sucking ability, exercise intolerance and a heart murmur.

She was astasia and the body temperature was 38.4°C. The jugular vein pulsation, tachycardia (heart rate 132 beats/min) and tachypnea (respiratory rate 40 breaths/min) were observed.

A maximum pansystolic heart murmur of grade III/VI was heard in the aortic valve area.

Hematological examination revealed a hematocrit of 37%, red blood cell count of $864 \times 10^4/\mu\text{l}$ and hemoglobin concentration of 13.2 g/100 ml. There was no polycythemia. The leucocyte count was 9,600 per μl . BUN and GOT were increased to 78 mg/100 ml and 78 K-U, respectively. However, the other hematological findings were normal.

The highly P wave in electrocardiograms (EKG) by A-B lead was 0.78 mV in amplitude which suggested a right atrium enlargement. The accentuated S wave in EKG by A-B lead was 2.44 mV in amplitude and suggested ventricular hypertrophy.

Cardiac catheterization was performed through the left jugular vein and carotid artery, but it was not possible to get the catheter into the pulmonary artery. Catheterization data were summarized in Table 1. Po_2 levels of the

left ventricle and carotid artery were lowered markedly (22.5 mmHg, 24.1 mmHg, respectively). Oxygen saturations of the left ventricle and carotid artery were very low (26.0%, 30.2%, respectively). The right ventricular pressure was elevated considerably (96.4 mmHg systolic / 0 mmHg diastolic).

Two-dimensional echocardiogram (2DE) was obtained using a right fourth intercostal scan. In 2DE, four chamber view was seen. Further cranial tilting of the transducer revealed the right ventricular outflow tract, ventricular septal defect and markedly dilated overriding aorta (Fig. 1). The pulmonary artery could not be visualized. Contrast echocardiogram (CE) documented a right-to-left cardiac shunting.

Fig. 1. Two-dimensional echocardiogram. The markedly dilated overriding aorta (AO) originated from right ventricular outflow tract (RVOT). Ventricular septal defect (VSD) is demonstrated clearly in the infundibular interventricular septum. RA; right atrium, AOV; aortic valve.

Table 1. Hemodynamic data

	Po_2 (mmHg)	Oxygen saturation (%)	Pressure data (mmHg) systolic/diastolic
Right ventricle	20.5	19.6	96.4/0
Left ventricle	22.5	26.0	98.2/0
Carotid artery	24.1	30.2	96.1/35.0

Fig. 2. Diagram summarizing the cardiovascular anomalies. The atria are normal in position. The ventricles demonstrate d-loop. The large solitary aorta (AO), with a trileaflet valve, originates from the left ventricle (LV) with mitral-aortic fibrous continuity, and overrides. Ventricular septal defect (VSD) is large and located underneath the aortic valve. The main pulmonary artery is atretic, and unrecognizable. In case 1, patent foramen ovale (PFO) is present. The left pulmonary duct artery (LPDA) branches off from the aorta and the right pulmonary duct artery (RPDA) branches off from the brachiocephalic artery. The bilateral pulmonary duct arteries supply the ipsilateral pulmonary arteries. In case 2, the LPDA branches off from the aorta and supplies the right pulmonary artery (RPA) and left pulmonary artery (LPA). In case 3, there is no connection between the atretic pulmonary artery (PA) and right ventricle (RV). The atretic PA, with a bicuspid valve, rises from the very small chamber which is located on the blind-ending muscular infundibulum. The PA shows post stenotic dilation. The LPDA branches off from the AO and supplies the LPA and RPA. A: case 1, B: case 2, C: case 3, RA; right atrium, LA; left atrium.

Fig. 3. Anterior view of the heart. The atria are normal in position. The apex of the heart is formed by the left ventricle (LV). A large solitary aorta (AO) overrides above the interventricular septum. The main pulmonary artery is atretic and unrecognizable. RA; right atrium, LA; left atrium, RV; right ventricle.

The calf continued to do poorly and died from hypoxemia and cardiac insufficiency at 11 days old, when her weight was 37.5 kg.

The cardiovascular findings were summarized in Fig. 2A. All viscera were in situs solitus and the ventricles demonstrated d-loop (Fig. 3). Ventricular septal defect was of a large perimembranous type and observed underneath the aortic valve. A large solitary arterial trunk (aorta) with a trileaflet valve originated from the left ventricle and overrode above the interventricular septum. There was a fibrous continuity between the aortic valve and mitral valve (Fig. 4). The main pulmonary artery was atretic and not recognized but an infundibular recess was present. The left pulmonary duct artery (LPDA) branched off from the aorta and the right pulmonary duct artery (RPDA) branched off from the brachiocephalic artery (Fig. 5). The bilateral pulmonary duct arteries supplied the ipsilateral pulmonary arteries. Patent foramen ovale was present with aneurysm of septum primum toward the left atrium, suggesting an increased right atrial pressure during life. On the other hand, the spleen was present and there was no malformation in the abdominal organ.

Case 2 (No. 001).

A 6-month-old female Holstein calf having cyanosis of the visible mucous membrane and exercise intolerance was examined. Body temperature was 39.4°C, heart rate 78 beats/min and respiratory rate 18 breaths/min. A maximum pansystolic heart murmur of grade III/VI was

Fig. 4. Internal view of the left ventricle (LV). There is a fibrous continuity between the aortic (AV) and mitral (MV) valves. Ventricular septal defect (VSD) is of a large perimembranous type, and underneath the aortic valve in location. AO; aorta.

Fig. 5. Ramification of the pulmonary artery. The left pulmonary duct artery (LPDA) branches off from the aorta (AO) and the right pulmonary duct artery (RPDA) branches off from the brachiocephalic artery. The bilateral pulmonary duct arteries supply the ipsilateral pulmonary arteries. LPA; left pulmonary artery, RPA; right pulmonary artery, RA; right atrium, LA; left atrium.

Fig. 6. Two-dimensional echocardiogram. The markedly dilated overriding aorta (AO) originated from right ventricular outflow tract (RVOT). RA; right atrium, RV; right ventricle.

Fig. 7. Anterior view of the heart. The atria are normal in position. The apex of the heart is formed by the left ventricle (LV). A large solitary aorta (AO) overrides above the interventricular septum. The main pulmonary artery is atretic and unrecognizable. RA; right atrium, LA; left atrium, RV; right ventricle.

Fig. 8. Internal view of the right ventricle (RV). There is a fibrous continuity between the aortic valve (AV) and mitral valve (MV) and also between the aortic valve and tricuspid valve (TV). A large infundibular ventricular septal defect (VSD) is observed underneath the aortic valve. The aorta overrides the VSD. AO; aorta.

heard in the aortic valve area.

Hematological examination revealed polycythemia with a hematocrit of 61%, red blood cell count of $1,880 \times 10^4/\mu\text{l}$ and hemoglobin concentration of 23.0 g/100 ml. The leucocyte count was 21,200 per μl . However, the other hematological findings were normal.

The accentuated S wave in EKG by A-B lead was 2.36 mV in amplitude and suggested ventricular hypertrophy. The phonocardiogram demonstrated a decrescendo-shaped murmur and a highly second sound.

2DE was obtained using a right fourth intercostal scan. In 2DE, four chamber view was seen. Further cranial tilting of the transducer revealed the right ventricular outflow tract, ventricular septal defect and markedly dilated overriding aorta (Fig. 6). The pulmonary artery could not be visualized. CE documented a right-to-left cardiac shunting.

She died at 8-month-old from hypoxemia and cardiac insufficiency.

The cardiovascular findings were summarized in Fig. 2B. All viscera were in situs solitus and the ventricles demonstrated d-loop (Fig. 7). A large infundibular ventricular septal defect was observed underneath the aortic valve. A large solitary arterial trunk (aorta) with a trileaflet valve originated from the left ventricle and overrode. There was a fibrous continuity between the aortic valve and mitral valve and also between the aortic valve and tricuspid valve (Fig. 8). The main pulmonary artery was atretic and not recognized. The LPDA branched off from the aorta and supplied the left and right pulmonary arteries (Fig. 9). On the other hand, the spleen was present and there was no malformation in the abdominal organ.

Case 3 (No. 1968)

A 9-month-old female Holstein calf was admitted to the

Fig. 9. Ramification of the pulmonary artery. The left pulmonary duct artery (LPDA) branches off from the aorta (AO) and supplies the right pulmonary artery (RPA) and left pulmonary artery (LPA). LA; left atrium.

Veterinary Medical Teaching Hospital, Rakuno Gakuen University, because of cyanosis of the visible mucous membrane, exercise intolerance and heart murmur.

Body temperature was 38.9°C, heart rate 63 beats/min. The jugular vein pulsation and tachypnea (respiratory rate 42 breaths/min) were observed.

A maximum pansystolic heart murmur of grade III/VI was heard in the tricuspid and pulmonary valve areas.

Hematological examination revealed polycythemia with a hematocrit of 53%, red blood cell count of $1,138 \times 10^4/\mu\text{l}$ and hemoglobin concentration of 18.0 g/100 ml. The leucocyte count was 3,800 per μl . However, the other hematological findings were normal.

The accentuated S and T waves in EKG by A-B lead were 2.25 mV and 1.20 mV, respectively in amplitude and suggested ventricular hypertrophy.

Po₂ level and oxygen saturation of the carotid artery were lowered markedly (28.0 mmHg, 43.0% respectively).

2DE was obtained from the right hemithorax. In 2DE, ventricular septal defect was seen clearly at the upper interventricular septum.

The cow continued to do poorly and died at 10-month-old from hypoxemia and cardiac insufficiency.

The cardiovascular findings were summarized in Fig. 2C. All viscera were in situs solitus and the ventricles demonstrated d-loop. The aorta and atretic pulmonary trunk mildly crossed each other as seen in the normally related great arteries (Fig. 10). A large perimembranous type ventricular septal defect was observed underneath the aortic valve. A large solitary arterial trunk (aorta) with a trileaflet valve originated from the left ventricle and overrode above the interventricular septum. There was a fibrous continuity between the aortic valve and mitral valve and also between the aortic valve and tricuspid valve (Fig. 11). The atretic pulmonary trunk, with a bicuspid valve, rose from a very small chamber which was located on the blind-ending muscular infundibulum (Figs. 12, 13). There was no connection between the atretic pulmonary trunk and the right ventricle. The LPDA branched off from the aorta and supplied both the left and right pulmonary arteries. On the other hand, the spleen was

Fig. 10 Anterior view of the heart. The atria are normal in position. The apex of the heart is formed by the left ventricle (LV). The aorta (AO) and pulmonary artery (PA) slightly crossed each other as seen in the normally related great arteries. The PA is atretic. RA; right atrium, LA; left atrium, RV; right ventricle.

Fig. 11. Internal view of the right ventricle. There is a fibrous continuity between the aortic valve (AV) and tricuspid valve (TV). A large perimembranous type ventricular septal defect (VSD) is observed underneath the aortic valve. AO; aorta.

Fig. 12. Internal view of the right ventricular outflow tract. There is a blind-ending muscular infundibulum (arrow). RV; right ventricle, VSD; ventricular septal defect.

present and there was no malformation in the abdominal organ.

TOF has been described by many researchers [2, 3, 5–13, 15–17, 19], and presently the diagnosis of TOF is possible with 2DE or M-mode echocardiography [2, 11, 15]. A ventricular septal defect, an overriding aorta, right ventricular hypertrophy were observed but the pulmonary artery could not be visualized with 2DE in three cases. We suspected them to have TOF with pulmonary atresia for these reasons. 2DE findings were confirmed with the autopsy findings in cases 1 and 2. However, we could not find atretic pulmonary artery and a small chamber which was located on the blind-ending muscular infundibulum in case 3 in 2DE, because the ultrasonic device we used was a linear scanning type and our 2DE techniques were incomplete. We considered that we should use the sector or convex type transducer and carry out a systematic 2DE approach for congenital heart disease. However, there may be practical difficulty in defining a severely hypoplastic pulmonary artery resembling a string [11], therefore we should have been applied angiocardiology for three cases.

Marked polycythemia developed in cases 2 and 3 as a result of arterial hypoxemia, but not in case 1. Case 1 died when 11-day-old, case 2 and 3 died at 8-month-old and 10-month-old, respectively. The patient with no polycythemia died earlier than those with polycythemia. We considered that it showed no reactions to arterial hypoxemia in case 1, therefore she died earlier than the other cases.

EKG showed an accentuated S wave in all cases, suggesting ventricular hypertrophy, which was confirmed by autopsy.

Pulmonary atresia describes the situation in which the

Fig. 13. Internal view of the atretic pulmonary artery (PA) and a very small chamber (SC). The atretic PA, with a bicuspid valve, rises from the SC which is located on the blind-ending muscular infundibulum. PV; pulmonary valve.

pulmonary circulation has no apparent connection with the ventricular mass. This could either be because the pulmonary valve is formed but imperforate or because the pulmonary trunk narrows down and ends blindly at the valve sinuses, with the ventricular myocardium interposing between the ventricular and arterial cavities. A further variation of the latter is when the entire pulmonary trunk segment, or even all of the intrapericardial pulmonary arteries, are absent [1]. All of the intrapericardial pulmonary arteries are completely absent in cases 1 and 2. These were categorized in truncus arteriosus, "truncus type IV" by using the classification of Collett and Edwards [4]. However, Becker and Anderson categorized them in the group of atresia with ventricular septum defect [1]. We agree with their opinion, so we treated them as TOF.

All of the intrapericardial pulmonary arteries are absent in cases 1 and 2. The pulmonary valve was formed and then perforated the small chamber but the infundibulum narrowed down at the valve sinuses in case 3. We think three cases to be rare.

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