

Cervical ectopia cordis in a calf: a case report

J. JEZEK, A. DOMANJKO PETRIC, T. PALLER, J. STARIC

Veterinary Faculty, University of Ljubljana, Ljubljana, Slovenia

ABSTRACT: A female Holstein Friesian calf with inferior cervical ectopia cordis was followed from the 16th day of age until euthanasia at the age of three months. The heart was located in the lower cervical region and was covered with skin. The calf was presented with normal heart and respiratory rate. A grade II/VI systolic murmur was audible at the heart base when the calf was checked for the first time, but this later disappeared. The ultrasonography revealed that the apex of the heart was directed cranio-ventrally with the left ventricle ventral and slightly to the right side while the right ventricle was dorsal. The results of complete blood count and biochemistry were inside reference intervals. At necropsy the heart was covered with normal pericardium and skin and displaced to the caudal part of the ventral cervical region, just cranial to an enlarged apertura thoracis cranialis. Additionally, the cranial lobes of the lungs were displaced through the enlarged thoracic aperture under the skin on the ventral part of the neck. In conclusion, we suspect that the deformed and enlarged cranial thoracic aperture enabled displacement of the heart and cranial lung lobes to the ventral neck region, because no other abnormalities of the heart and vessels were observed. This is the first case of cervical ectopia cordis reported in Slovenia and this article provides data regarding different aspects of the case (clinics, laboratory, pathomorphology) while also discussing previously published cases of ectopia cordis in cattle.

Keywords: ectopic heart; bovine; congenital malformation; displacement of heart

Ectopia cordis is a rare congenital malformation characterised by a complete or partial displacement of the heart outside the thoracic cavity. It may be subdivided into three types; cervical, sternal (thoracic) and abdominal. Based on the position of the heart in the cervical region, the cervical type may in turn be divided into two further subtypes: superior and inferior. The inferior cervical type is the most common reported type of ectopia cordis and the most compatible with life (Bowen and Adrian 1962; Windberger et al. 1992). In the sternal type the heart is partially or completely displaced through of a defect in the sternum or ribs. The abdominal type is the least frequent. Here, the heart lies in the epigastric, lumbar or umbilical region with the great vessels and is displaced through a defect in the diaphragm. In some cases the heart herniates through a split in the abdominal wall. The ectopic heart may lack skin coverage and in some cases also the pericardium (Bowen and Adrian 1962).

The exact pathogenesis of ectopia cordis is unknown, and most cases are sporadic. The incidence in humans is reported to be 5.5–7.9/million live births

(Morales et al. 2000). Ectopia cordis was reported also in pigs (Freeman and McGovern 1984; Sabolic et al. 2006) and goats (Ramadan and Abdin-Bey 1993). In a 14-year study Gopal et al. (1986) found ectopia cordis cervicalis in 10 out of 36 calves with cardiac defects.

The aim of this paper is to describe clinical and laboratory findings and morphological abnormalities in a female Holstein Friesian calf with inferior cervical ectopia cordis and to review previously reported cases.

Case description

History and clinical findings. The gestation length and parturition of the female Holstein Friesian calf were normal but after birth the owner noticed enlargement and pulsation on the neck of the calf. The calf was 16 days old when it was examined for the first time. It was bright, alert and responsive with normal appetite. The heart was located in the lower cervical region and was covered with skin. The heart rate was 94/min and respiration rate was 40/min. Mucus membranes were

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Figure 1. The calf with cervical ectopia cordis

slightly cyanotic. Continuous low intensity murmur grade II/VI was audible on the heart base. No abnormal sounds were found by lung auscultation.

The calf was re-examined 17 days later when it was 33 days old. The owner noticed lower weight gain and that the calf often coughed while eating forage. The heart rate was 95/min and respiration rate was 40/min, the CRT was 2–3 s. Low intensity systolic murmur (II–III/VI) was still audible at the heart base.

At the age of 2.5 months the calf was admitted to the Clinic for Ruminants for further observation. At that time the general appearance of the calf was normal. Vital signs were within normal ranges ($T = 38.9\text{ }^{\circ}\text{C}$, $P = 62/\text{min}$, $R = 48/\text{min}$, $R_c = 7/5\text{ min}$). The mucus membranes were slightly cyanotic; CRT was 2–3 s. Heart auscultation revealed no abnormal heart sounds. After moderate exercise we observed increased respiratory rate and slight mixed dyspnoea.

Laboratory findings. At the age of 16 days haematological values were the following: RBC ($8.05 \times 10^{12}/\text{l}$), Hb (109 g/l), Ht (0.33 l/l) and MCV (41 fl). Haematological values at the age of 77 days were, RBC ($10.56 \times 10^{12}/\text{l}$), Hb (123 g/l), Ht (0.37 l/l) and MCV (35 fl). The values of other haematological variables changed only negligibly between samplings. Biochemical examination at the age of 77 days revealed, CK (257 IU/l), AP (163 IU/l), GGT (14 IU/l), total serum protein (63.6 g/l), urea 4.03 mmol/l, Ca (2.62 mmol/l), iP (2.41 mmol/l), Mg (1.07 mmol/l), and serum iron (22.6 $\mu\text{mol/l}$).

Electrocardiographic and ultrasound examination. The base-apex electrocardiogram showed no abnormalities.

Ultrasonography revealed that the apex of the heart was directed cranio-ventrally, the left ventricle was

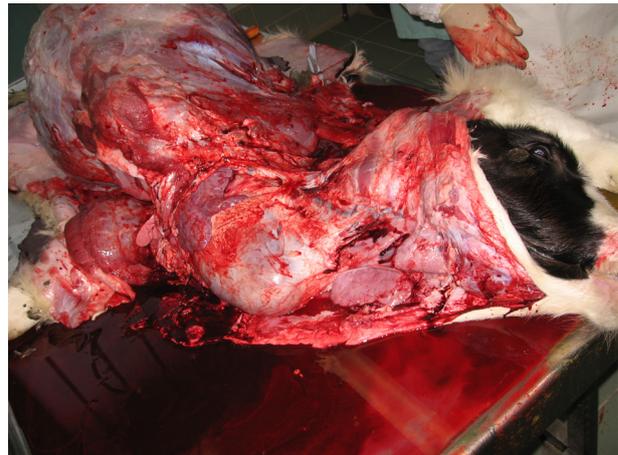


Figure 2. Pathomorphology; the heart covered with pericardium, atelectatic cranial lung

located ventrally slightly to the right and the right ventricle was dorsal. The right ventricle was slightly dilated. The aorta and truncus pulmonalis were directed caudally, and positioned normally. Both atria were of appropriate size. Blood flows were normal except for trivial insufficiency of the aortic valve.

Pathomorphological findings. The calf was euthanised and sent for necropsy. The heart was positioned under the skin on the ventral part of the neck. The apex was directed cranio-ventrally, the right side of the heart was oriented laterally to the left and the left ventricle was oriented laterally to



Figure 3. Enlarged cranial thoracic aperture

the right (torsion for 90° to the left). The heart was covered with normal pericardium; within the pericardial sac 1.5 dcl of yellow transparent fluid was found. The atria and both ventricles were dilated. Cranial thoracic aperture was oversized (11 cm × 11 cm) and the sternum was cleft cranially. The cranial lobes of the lungs were atelectatic and displaced through the enlarged cranial thoracic aperture under the skin on the ventral part of the neck. The liver and kidneys were hyperaemic. Mucus membranes were cyanotic. No other anatomical abnormalities were observed.

DISCUSSION AND CONCLUSIONS

The most frequently reported types of ectopia cordis in cattle are cervical (82%), pectoral (14%) and abdominal (3%). The cervical type is the most compatible with life (Hiraga and Abe 1986; West and Payne-Johnson 1987) while calves with the other types died before or early after birth (Eroksuz et al. 1998; Schulze et al. 2006). Ectopic heart cases can be complicated by different kinds of cardiac and non-cardiac malformations (Hiraga and Abe 1986; Hiraga et al. 1993; Eroksuz et al. 1998. West

Table 1. Review of previous reports of ectopia cordis in calves

Reference	Breed, sex	Age	Form of ectopia	Heart malformations	Further findings
Bowen and Adrian 1962	SH, F	22 months	C + P	DA, AV, ductus Botalli	SM, abnormally shaped lungs
	SH, F	3 days	C + P	PFO	
Kurtz and Ellery 1969	HF, F	fetus (late stage 3 rd trimester)	S + P	dilated atria	SM
Fiedler and Seidler 1974	A, F	5 weeks	C + P	DA, PFO, AV, dilated right ventricle, ductus Botalli	SM
Hiraga and Abe 1986	HF, F	29 days	C + P	DA	SM, torticollis
	HF, F	20 h	C + P	DA	SM, cleft palate
	HF, M	1 day	C + P	DA	SM, torticollis
	HF, M	312 days	C + P	DA, ductus Botalli	SM
	HF, F	1 day	C + P	DA	SM, torticollis
	HF, F	5 min	C + P	DA	SM, torticollis, cleft palate, diaphragmatic hernia
	HF, M	10 min	C + P	DA	SM, torticollis
	HF, M	3 min	C + P	DA	SM, torticollis
West and Payne-Johnson 1987	HF, F	8 months	C + P	PFO, interventricular septal defect	SM, abnormally shaped lungs
	HR, M	14 months	C + P	PFO	
Windberger et al. 1992	FV, M	21 months	C + P	AV, myocardial hypertrophy	
Hiraga et al. 1993	H, M	0	C + P	DA	SM, torticollis, cleft palate, bilateral cryptorchidism
	H, F	1 h	C-S + P	enlarged heart (750 g)	SM, torticollis partial cleft palate
Eroksuz et al. 1998	H, F	30 h	S + P	dilated atria	SM
Schulze et al. 2006	GH, M	0	S + P		thoracoschisis
Shirian et al. 2010	HF, M	10 min	C + P	DA	SM, torticollis
Hamali and Ashrafi-helan 2010	H, M	2 min	C + P	rounded shape of the heart	
Onda et al. 2011	H, F	3 years, 4 months	C + P	dysplasia of the tricuspid valve	SM
Frackowiak et al. 2014	L, F	newborn	S-	DA, PFO, AV	SM

HF = Holstein Friesian, H = Holstein, GH = German Holstein, A = Angler, HR = Hereford, L = Limousin, SH = Shorthorn, M = male, F = female, C = ectopia cordis cervicalis, S = ectopia cordis sternalis (pectoralis), P = pericardium present, DA = double apex, PFO = patent foramen ovale, AV = abnormalities of great vessels, SM = sternum malformation

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and Payne-Johnson (1987) observed tachycardia, increased respiratory rate and a positive jugular pulse in a three-month old calf with cervical ectopia cordis. They found a patent foramen ovale and a ventricular septal defect at *post mortem*.

Most cases of ectopic heart were reported in the Holstein Friesian breed (Hiraga and Abe 1986; Hiraga et al. 1993; Schulze et al. 2006; Onda et al. 2011). The condition has also been reported in Angus, Shorthorn, Hereford, Guernsey, Simmental, Limousin and mixed breed animals (Hughes 1934; Bowen and Adrian 1962; Gopal et al. 1986; Windberger et al. 1992; Frackowiak et al. 2014).

The clinical signs observed in our case, such as coughing, slight dyspnoea after exercise and cyanotic mucus membranes were in accordance with the findings of other reported cases (Bowen and Adrian 1962; Hiraga and Abe 1986; Windberger et al. 1992). Cough may result from the pressure of the heart on the trachea when the calf lowers the head. Dyspnoea could be a consequence of atelectatic cranial lung lobes in our case.

A comparison of haematological findings between the first and the second sampling established that RBC number, and Hb and Ht values increased and MCV values decreased. The increased RBC count, and Hb and Ht concentrations are partly age-related, while the values could also partly reflect the response of the organism to lower lung capacity due to atelectasis of the cranial lung lobes. However, the values were still inside reference intervals for calves (Jezek et al. 2011). This explains the clinically observed cyanosis and dyspnoea after exercise. Biochemistry results were within reference values for this age (Jezek et al. 2006). Windberger et al. (1992) also reported normal haematological and biochemical values in a calf with ectopia cordis cervicalis at the age of three weeks. Onda et al. (2011) observed no abnormalities with respect to complete blood count, blood biochemistry and blood gas analysis. No other reports describing haematological and biochemical results were found in the literature.

Ultrasound examination revealed no anatomical abnormalities of the heart except trivial insufficiency of the aortic valve. With respect to instances of cardiac malformation in the literature, Onda et al. (2011) reported that in the case of a five-day old calf with cervical ectopia cordis ultrasonography revealed dysplasia of the tricuspid valve. No other reports of ultrasound findings were found in the literature regarding ectopia cordis.

As in other reported cases of cervical ectopia cordis (Bowen and Adrian 1962; Fiedler and Seidler 1974; Hiraga and Abe 1986; Windberger et al. 1992), the heart was located in the cervical area and was covered with normal skin and pericardium. In cases of thoracic ectopic heart the heart protruded through the defect in the sternum and was not covered with skin (Kurtz and Ellery 1969; Hiraga et al. 1993; Eroksuz et al. 1998; Schulze et al. 2006; Frackowiak et al. 2014). Hiraga and Abbe (1986) observed no abnormalities in the heart itself except for a double apex in a cervical ectopia cordis case. In our case the heart was normal except for slight right ventricular dilatation, which was detected ultrasonographically. This was most likely a consequence of increased pulmonary resistance due to atelectatic cranial lobes. Bowen and Adrian (1962) observed double apex, dilatation of the right ventricle and a larger than normal heart. Windberger et al. (1992) observed myocardial hypertrophy of both ventricles in a 21-month old calf with a cervical ectopic heart, which in the authors' opinion, was caused by the altered anatomical situation of the great vessels. Also, in a case of a thoracic ectopic heart, no abnormalities of the heart were observed (Schulze et al. 2006). Some authors have observed anomalies in the great vessels derived from the heart, which may be attributed to developmental failure in the early embryological stage (Fiedler and Seidler 1974; Windberger et al. 1992; Hiraga et al. 1993). The cranial thoracic aperture in our calf was much wider than normal, which is in agreement with other reports (Bowen and Adrian 1962; Hiraga and Abbe 1986). The cranial lobes of the lungs protruded through the enlarged thoracic aperture under the skin, on the ventral part of the neck. This most likely caused atelectasis, similarly to what was observed in Case 3 reported by Hiraga and Abbe (1986). In Case 1 reported by Hiraga et al. (1993) the cranial part of the right lung lobe protruded into the pericardial cavity while the sternum was cleft cranially and was fused only at the xyphoid process. In our case only the presternum was cleft. Windberger et al. (1992) observed emphysema and atelectasis combined with follicular peribronchitis in patches throughout the lung.

Liver and kidney hyperaemia were observed in our case, most likely due to congestion. Hyperaemia of various internal organs was also observed in another case of ectopia cordis (Schulze et al. 2006).

On the basis of the findings in this case and reports from other authors, we suspect that deformed cranial thoracic aperture was the cause of the ec-

topa cordis cervicalis, because no other abnormalities of the heart and vessels were found.

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Corresponding Author:

Jozica Jezek, University in Ljubljana, Veterinary Faculty, Clinic for Ruminants with Ambulatory Clinic, Cesta v Mestni Log 47, 1000 Ljubljana, Slovenia
E-mail: jozica.jezek@vf.uni-lj.si