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An unusual case of thoracic ectopia cordis in a Toggenburg Goat and its three-dimensional images constructed with X-ray computed tomography

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Abstract

A two-day-old female Toggenburg goat with thoracic ectopia cordis (EC) was diagnosed via radiography and computed tomography. The goat was born with EC, defects of the sternum and a supra-umbilical abdominal wall, but without the presence of Cantrell's syndrome. Necropsy and histopathological findings indicated the affected kid had malformation of the heart with an enlarged left ventricle. The findings showed the heart (9 × 5 × 5 cm) stayed outside the thorax, and was covered by a semitransparent membrane. This report is the first to describe a case of thoracic EC in a goat whose sternum was not developed fully and was not connected to the ribs. It is also the first paper to describe three-dimensional images of this condition constructed from computed tomography scans.

Key Words: computed tomography, congenital malformation, ectopia cordis

Ectopia cordis is defined by the presence of a heart located partly or fully outside the thorax^{3,8)}. According to the position of the heart, ectopia cordis can be categorized into five types: cervical, cervicothoracic, thoracic, thoracoabdominal, and abdominal⁸⁾.

According to the data collected from a children's hospital in Canada, the prevalence of ectopia cordis in humans ranges from 5.5 to 7.9 per million live births^{5,6)}. Ectopia cordis in animals

have been reported in cattle, goats^{8,11,13)} and horses^{2,3)}, but cases in goats have not been widely reported and there have been no reports of cases in which the sternum has not fully fused with the ribs. Many dairy goat breeds, such as Alpine, Nubian, Saanen, Toggenburg, and Lammancha, have been well established¹⁰⁾. Toggenburgs, a friendly and gentle breed with an average of 3 to 5 Kg milk production per day¹⁰⁾, is popular in Taiwan. To date, very few reports mentioned the

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Fig. 1. The membrane was semitransparent. Fluid was seen inside. The umbilicus was located on the antelope side of the membrane and on the skin of abdomen. The arrow indicates the position of umbilicus.

congenital problems in Toggenburgs and the published ectopia cordis cases in goat were not this breed^{9,11,13}.

There are also no reports of using X-ray computed tomography to assist with the diagnosis of thoracic ectopia cordis in goats. This report describes a rare case of thoracic ectopia cordis in a Toggenburg goat, in which the sternum did not fuse completely; and the three-dimensional structure of the thorax in the affected goat, which was constructed by computed tomography.

History and clinical signs: A two-day-old (D day) female Toggenburg goat was sent to a veterinary hospital because she was born with her heart outside of the thorax, covered by a semitransparent membrane (Fig. 1). The heart was located near the border of her thorax and abdomen. The goat was active and had a good appetite, although a mild weakness was noted. The goat weighed 3.5 kg and her body temperature was 37.6°C. The goat's heart rate was 210 beats per minute (normal range: 140 to 180 beats per minute), and her respiratory rate was 150 times per minute (normal range: 20 to 40 times per minute¹⁰). Capillary refill time was less than 1.5 seconds, after applying finger pressure to the goat's gums. Examination of the goat's scruff and gums showed six to eight percent dehydration. The gum's color was pink. The heart was covered by a semitransparent membrane, located around

the junction site of thorax and abdomen. There was fluid inside the membrane. The umbilicus was located at the rear site of the membrane (Fig. 1).

Hematological examinations: Hematological parameters of the affected goat were evaluated at D+1 days after admission (Table 1). The goat showed signs of dehydration and anemia, with elevated liver enzymes.

Radiographic and ultrasonic examinations: From the images, the border of the heart could not be identified under the right lateral-recumbent due to a fast heart beat. The lung did not expand well. The terminal sites of ribs did not connect to the sternum. There was no severe back blood flow during Doppler ultrasound evaluation (data not shown).

Computed tomography scan: The goat was placed in the dorsal-ventral recumbent position after the animal was sedated by given propofol (6 mg/kg *i.v.*) and a contrast agent (0.2 mL/kg of Magnevistag[®], Bayer Pharma AG, *i.v.*). Then, the goat was taken images with a CT machine (GE HiSpeed DX/I, GE, USA), based on the protocol: slice thickness = 2 mm, KV = 120 KVp, mA = 150 mA, rotor per second = 1 sec, pitch = 1.25 and helical scan. A pleural effusion line could be observed on the images (Fig. 2). The heart was located almost fully outside the thorax; however, the thoracic diaphragm was normal. The three-

Table 1. Hematological and blood biochemical values of the affected goat with thoracic ectopia cordis

Items ^a	Unit	D+1 day	References ^{4,6)}
RBC	10 ⁶ /μL	7.38 ↓	10.1–16.7
Hb	g/dL	9.5 ↑	7.5–9.3
PCV	%	27.5	21.7–28.5
MCV	fL	37.3 ↑	15–24.6
MCH	Pg	12.9 ↑	5.2–8.0
MCHC	g/dL	34.5	30–37.8
PLT	10 ³ /μL	219 ↓	300–600
WBC	10 ³ /μL	3.51 ↓	4.58–10.46
Band	(%) n/μL	0	0
Neu.	(%) n/μL	(64.1 ↑) 2,250 ↓	(37.3–73.1) 2,636–5,497
Eos.	(%) n/μL	(0.3) 10 ↓	(0.7) 25
Baso.	(%) n/μL	(3.4) 120 ↑	(0.2) 7
Lymph.	(%) n/μL	(32.2) 1,130	(26.4–56.2) 927–1,973
Mono.	(%) n/μL	(0) 0 ↓	(0.7–3.3) 25–116
ALT	U/L	0	–
AST	U/L	108 ↓	167–513
ALKP	U/L	674 ↑	87.7 ± 29.2
BUN	mg/dL	58 ↑	10–20
Creatine	mg/dL	1.0	1.0–1.8
T-Prot	g/dL	3.8 ↓	6.4–7.0
T-Chol	mg/dL	20 ↓	80–130
T-Bili	mg/dL	0.3 ↑	0–0.1
Glucose	g/dL	88 ↑	50–75
Albumin	g/dL	1.4 ↓	2.7–3.9

^aPLT platelets, RBCs red blood cells, PCV packed cell volume, Hb hemoglobin, MCV mean corpuscular volume, MCH mean corpuscular hemoglobin, MCHC mean corpuscular hemoglobin concentration, WBCs white blood cells, Baso basophils, Eos eosinophils, Lymph lymphocytes, Mono monocytes, Neu neutrophils, Band band neutrophils. ALT alanine aminotransferase, AST aspartate aminotransferase, ALKP alkaline phosphate, BUN blood urea nitrogen, T-Chol total cholesterol, T-Prot total protein, T-Bili Total bilirubin
 -: No data.

dimensional image revealed an under-developed sternum, which resulted in the ribs being unable to contact the sternum (Fig. 3). 3D imaging combining the soft tissues showed that the heart was located from the 3rd to the 8th rib (Fig. 4). The CT images at the same places in Fig. 4 were shown in Fig. 2.

Fine needle findings: Cytospin technique was used to evaluate the content of the pleural effusion. Cells collected from the effusion were stained by Lui's stain. Some fibrin-like materials, segment non-regenerated neutrophil and some

cells were observed (data not shown).

Patho-anatomical and histopathological findings: The goat died at D+3 days and a necropsy was performed. Patho-anatomical studies showed that the goat had hydrothorax. The ribs were not connected to the sternum and the areas by the sides of the ribs were replaced by soft tissues. The heart was located around the junction of the thorax and abdomen. The walls of the thorax and abdomen were not defective. The anteroapical of the mass was near the site of the umbilici. The sternum was not fused fully. The

diaphragm was normal. The affected goat did not show the five signs of pentalogy of Cantrell⁷. The patho-anatomical and histopathological findings from the heart, lung, liver, kidney, spleen and stomach were listed in Table 2.

Discussion

Although some publications have mentioned goat ectopia cordis based on pathological and ultrasonic findings^{9,11,13}, this report is the first on 3D thorax images of a Toggenburg goat affected

by thoracic ectopia cordis.

Congenital thoracic ectopia cordis is caused by the incomplete fusion of the body cavity, especially in the abdominal wall, during development of the fetus. The impaired process results in the incorrect position of the heart after the fetal development³. Most reports on ectopia cordis have been focused on humans and cattle³, and there is little in terms of descriptions of thoracic ectopia cordis in goats. The prevalence of thoracic ectopia cordis varies among species. Sixty percent of all human cases and 14% of all reported cattle cases were identified as the

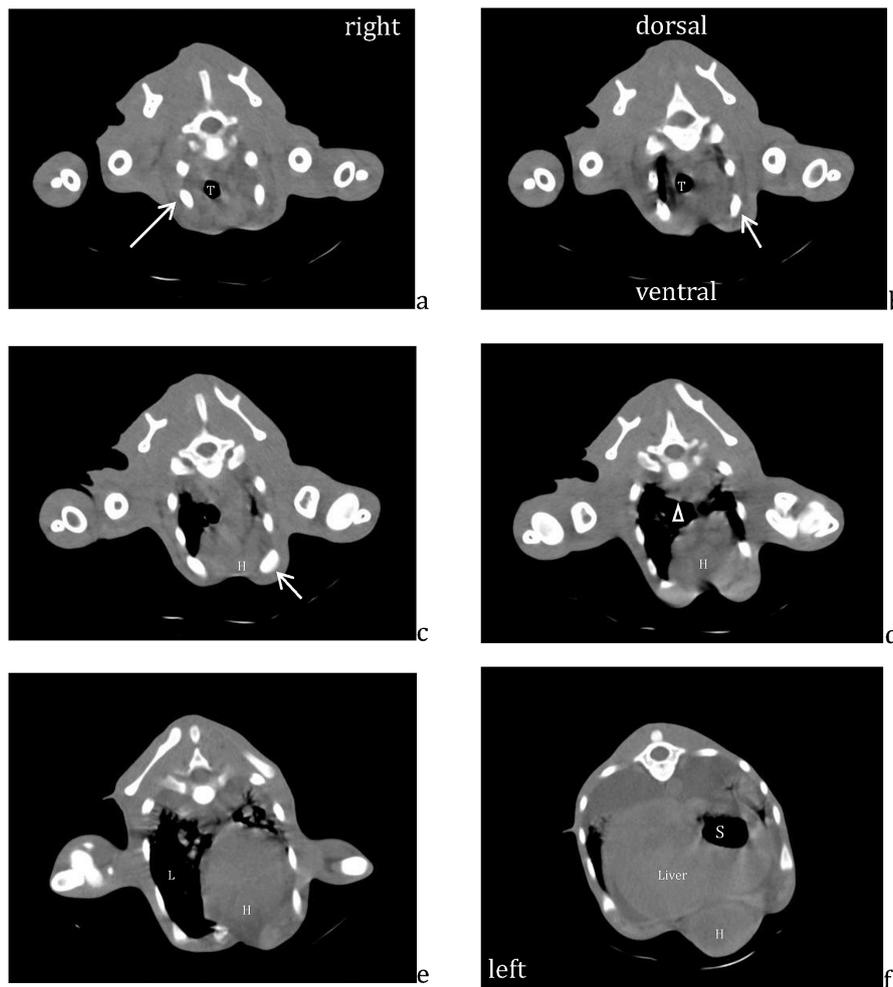


Fig. 2. The computed tomographic images from locations a, b, c, d, e and f show the change the structure of organs in thorax. The goat was placed in the dorsal-ventral recumbent position but the images were 180 degree rotation. The space of lung was insufficient, which was occupied by pleural effusion. The heart was located outside the thorax. The sternum was disappeared and did not fuse properly at the abdominal midline. The CT images were used to build the following Fig. 3 and Fig. 4. The corresponding locations were listed in Fig. 4. *H* heart, *L* lung, *S* stomach, *T* trachea, *arrow arrowhead* pleural effusion line.

thoracic type. There was a reported case of thoracic ectopia cordis in a cattle with developed sternebrae³⁾. However, the affected goat in this study had 12 pairs of ribs and underdeveloped sternebrae which was not attached to its ribs.

Congenital deficiencies such as omphalocele and amniotic band syndrome may confuse the diagnosis of ectopia cordis^{8,12)}. Omphalocele is caused by hypoplasia of the abdomen wall. Amniotic band syndrome is caused by the broken amniotic membrane, while the chorionic membrane is still intact. Broken amniotic membrane can cause

defects of organs or limb trunks to grow symmetrically. Congenital ectopia cordis would be related to amniotic band syndrome^{8,12)}. In this condition, the band interferes and presses the heart, forcing it to move to the abdomen and results in ectopia cordia. Some reports also described a situation in which the heart was

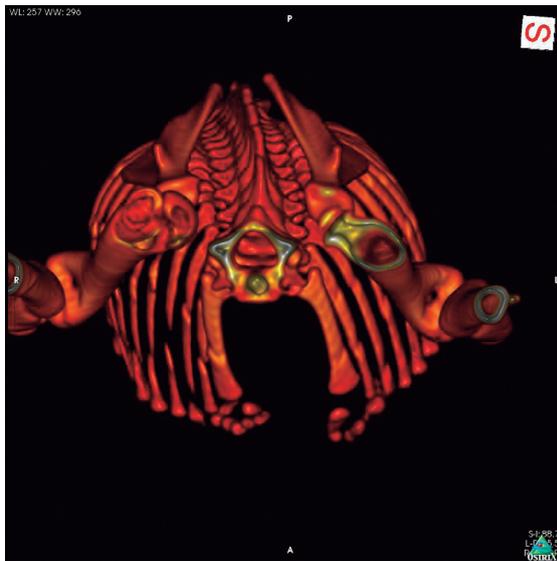


Fig. 3. Three-dimensional image constructed using bones only in OsiriX imaging software V6.0. From the head side, a sternum was not developed and the ribs were not able to connect to the sternum. The other part of the thorax was developed properly.

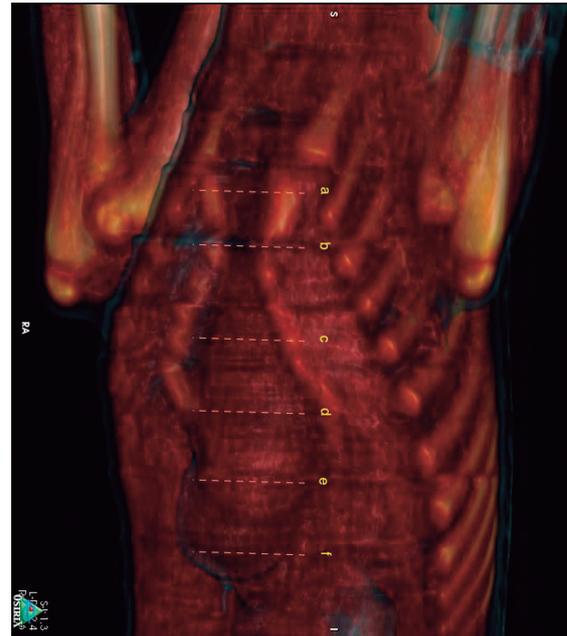


Fig. 4. Three-dimensional image was constructed by mixing the bone structure and soft tissues in OsiriX imaging software V6.0 in Fig. 4. The CT images were relocated based on the dorsal-ventral and the left-right sides. The abnormal heart was located from the 3rd rib to the 8th rib. The sternum was disappeared and the ribs were not connected. The CT images at the locations of a, b, c, d, e, and f were shown in Fig. 2.

Table 2. Patho-anatomical and histopathological findings of the affected goat with thoracic ectopia cordis

Organs	Patho-anatomical findings	Histopathological findings
Lung	Marble-like surface; hemorrhagic or bleeding spots on the surface	Atelectasis
Heart	Heart located around the junction of the thorax and abdomen; 9 × 5 × 5 cm; fibrin-like materials on the surface; left ventricle enlargement	Thickened and hemorrhagic pericardium; neutrophils infiltration
Liver	Fibrin-like materials on the surface; gross distortion caused by the ribs pressing on	Congestion
Kidney	Not well developed; unclear border between cortex and media	Normal
Spleen	Pale	Normal
Stomach	Milk clots	Normal

unable to move to the correct position because it was bound by umbilici^{8,12}). Defects of cattle's thorax and sternum have been well discussed but disappearance of the sternum has not been reported³). Most of the congenital ectopia cordis cases had deficiencies inside the heart¹) or had abnormal pericardial cavity³). The affected goat in this report had a distorted heart which was caused by the pressure of its ribs. The length of the long axis of the heart was nine cm but there was not a vivid blood back-flow in ultrasonic photogram. The patho-anatomical findings did not show any abnormality inside the heart. The heart was contained inside the pericardial membrane, covered by a semitransparent membrane. The tissues inside the heart were normal, and all organs appeared normal except the size of the heart. The sternum was not present, and no any unsymmetrically limbs were found.

In summary, this case was the first case of thoracic ectopia cordis in goats in which the sternum had not fused completely, and the first time when the diagnosis was confirmed by computed tomographic and patho-anatomical analyses. The case showed that the congenital defect was only limited in the thorax.

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