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Cervico-pectoral Ectopia Cordis in Two Holstein Calves

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Abstract. Two Holstein calves affected with cervico-pectoral ectopia cordis, a male (No. 1) delivered stillborn and a female (No. 2) died 1 hour after birth, were examined macroscopically and radiographically to assess the severity and elucidate the teratogenesis of the anomaly. The heart of one calf was covered by the intact pericardium and skin and displaced to the caudalmost portion of the ventrocervical region, just cranial to an enlarged thoracic inlet. The cranial vena cava and the vena cava azygos were duplicated. The sternum was bilaterally divided into two parts fused only at the xiphoid process and was semicircular. The heart of the other calf, covered solely by the pericardium, was exposed ventrally from an oval opening in the body wall just cranial to a defective Y-shaped sternum. In both calves, the arterial branching pattern from the aortic arch was intermediate between the patterns of the dog and pig, and in each case a single pulmonary vein emptied into the left atrium. Pronounced or slight torticollis, cleft palate, and abnormalities in the urogenital system were also found. From an embryological perspective, cervical and pectoral ectopia cordis have a common pathogenesis; the cervical type occurs at a slightly earlier fetal stage than the pectoral type.

Key words: Cattle; congenital malformation; ectopia cordis; pathogenesis.

Ectopia cordis (EC), a condition defined as the abnormal position of the heart anywhere outside the thoracic cavity, is subdivided into three types according to the site of displacement: cervical, pectoral, and abdominal. In cattle, the cervical type is the most prevalent. A rare intermediate type of EC, partially cervical and partially pectoral, has been documented in only one bovine case.

Two examples of the intermediate type were presented at our laboratory recently. Necropsy of two neonatal calves revealed EC characterized by the relocation of the heart between the cervical region and the pectoral region. To elucidate possible embryological implications in the teratogenesis of EC, the gross anatomy of these animals was compared with that of the calves with the other types of EC defects.

Materials and Methods

Within a 34-month period, two calves with severe ectopia cordis (EC) were born on separate farms in Hokkaido (northern Japan). Death was ascribed to severe complications from the EC. Both calves were brought dead to the laboratory. Calf No. 1, a Holstein male, was delivered stillborn by Caesarean section on 21 April 1987 and was the third offspring of a multiparous dam that had previously borne two viable, normal calves. Calf No. 2, a Holstein heifer born 2 February 1990, died 1 hour after birth; the dam had a history of seven normal deliveries.

In preparation for postmortem examination, calf No. 1 was perfused with 2% formalin via the right femoral artery, and calf No. 2 was perfused via the left common carotid artery. The skeletal systems were macerated, and the ster-nums were radiographed with soft x rays (Softex K-2, Softex Co., Tokyo, Japan).

Results

Calf No. 1

This calf was completely developed and weighed 50 kg (usual size for a male neonate Holstein). The skin was intact, and the hair coat was normal. However, pronounced torticollis to the right was evident, and a swelling was apparent in the caudalmost cervical region just a few centimeters cranial to the enlarged thoracic inlet (Fig. 1).

The left and right sternoccephalic muscles, originating bilaterally from the sternum, fused with the brachiocephalic muscles. The fused pairs of muscles were thin and fatty and cranially diverged at the swelling in the caudal cervical region (Fig. 1). The fibrous pericardium was connected to the mandibles by several broad, flat ligaments. The sternum was abnormally semicircular and was attached to the fibrous pericardium at the cranial margin by wide, thin sheetlike ligaments. The pericardial cavity, although intact, was extremely large (Fig. 2).

The heart was round, weighed 350 g, was enveloped in the pericardium, and was situated in the swollen part of the ventrocervical region underneath the subcutaneous layer and skin. Inside the spacious pericardial cavity, the heart was characterized by a double apex, which pointed either cranially or caudally, depending on the general position of the animal (Fig. 2).
In contrast to the apex, however, the base of the heart was somewhat anchored by the great vessels just beneath the VIth cervical vertebra. The lungs comprised two left and six right lobes, with the cranialmost two of the right lobes protruding into the pericardial cavity (Fig. 2).

Immediately after the aorta emerged from the pericardium, the brachiocephalic trunk and then the left subclavian artery branched off. The brachiocephalic trunk branched from a common site into the left and right common carotid arteries and the right subclavian artery (Fig. 3). The cranial vena cava and the vena azigos were duplicated. Because of the extrapericardial displacement of the heart, the thoracic portion of the caudal vena cava was considerably elongated, but this did not result in any noticeable diminution of the diameter of the vessel. A single pulmonary vein emptied into the left atrium (Fig. 3).

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Fig. 2. Heart, pericardial cavity; calf No. 1. The heart, its apex pointing caudally, and two lobes of the right lung (arrowheads) occupy the intact pericardial cavity. The fibrous pericardium is connected with the sternum by a wide ligament (*) and several thin ligaments. Bar = 5 cm.

Fig. 3. Right dorsal view, heart and great vessels; calf No. 1. The left subclavian artery (Ls) arises independently from the aortic arch, and the brachiocephalic trunk branches into the left (L) and the right (R) common carotid and the right subclavian (Rs) arteries at a common site along the trunk. The left (La) and right (Ra) azigos veins empty into the left and right cranial vena cavae, respectively. Vp = common pulmonary vein. Bar = 5 cm.
The sternum parted craniocaudally into two equal pieces and was fused only at the xypophoid process. Consequently, the gross appearance of the sternum was semicircular. The bone comprised 19 irregular sternebrae, as determined radiographically (Fig. 4, D). Several thin, membranous ligaments bridged the space between the left and right pieces of the sternum and tautly connected the pericardium to the defective sternum (Fig. 2).

In addition to the primary deformities in the neck and chest of the animal, cleft palate and bilateral cryptorchidism were present.

Calf No. 2

This calf was a fully developed female Holstein born at full term and weighed 41 kg. However, the heart, protected solely by the pericardium, was exposed ventrally from an oval opening (7 x 5 cm) in the body wall at the cervico-pectoral region (Fig. 5).

The bilateral sternoccephalic muscles originated cranially from the right tip of the abnormal Y-shaped sternum (Figs. 6, 7). The left m. pectoral descendens originated from the left margin of the dermal hole on the cranial side of the opening (Fig. 7). The fibrous pericardium enclosing the heart was attached to the rim of the dermal opening (Figs. 5, 6). The pericardial cavity was imperfectly formed because of partial adhesion between the parietal and visceral pericardia (Fig. 8). No ligaments extended from the pericardium.

The heart was grossly enlarged, with a weight of 750 g. It protruded through the oval opening in the body and was suspended solely by the great vessels (Figs. 5, 6), and tilted dorsocaudally in such a manner as to direct the apex cranioventrally. The lung appeared normal.

The branching pattern of vessels from the arch of the aorta in calf No. 2 was comparable to that found in calf No. 1, with the notable exception of a shorter interval between the sites of origin of the brachiocephalic trunk and the left subclavian artery (Fig. 8). Only one pulmonary vein emptied into the left atrium. The cranial vena cava was normal, but a right vena azygos, rather than the normal left one, was present.

The sternum was divided longitudinally into two parts that extended bilaterally from the manubrium to the middle of the body and formed an obvious Y-shape. The cranial margin of the bone had a smoothly finished edge and formed the caudalmost half of the opening in the skin through which the heart was exposed (Fig. 7). The Y-shaped sternum was determined by radiographic examination to be formed by 10 irregular sternebrae (Fig. 4, E).

Additional defects included slight torticollis, a partial cleft palate, and an abnormally large clitoris.

Fig. 5. Head, neck, ventral thorax; calf No. 2. The extremely enlarged heart protrudes ventrally from the thorax and is suspended only by the great vessels. Bar = 10 cm.

Discussion

The peculiar anatomical defects present in the two calves were compared with anomalies associated with cervical and pectoral ectopia cordis (EC) seen in nine other bovine cases previously investigated in our laboratory. These defects of the heart, artery, veins, and sternum and other miscellaneous abnormalities are summarized in Table 1. In contrast to reported cases of bovine EC, the displacement of the heart in these two calves took a middle position, between those of cervical and pectoral EC. A number of complications found in calf No. 1 are common in bovine cervical EC, including the double apex of the heart, the ligaments connecting the mandibles and the sternum, and the duplication of the cranial vena cava and the vena azygos. The anomalies of calf No. 2 were closer to those seen in bovine pectoral EC. A dermal opening exposing the heart is common in cases of EC involving the chest.

In most reports of bovine cervical EC, the apex of the heart pointed in a cranial direction, and the base tilted caudally. In calf No. 1, a large pericardial cavity permitted a range of movement of the apex. This unconventional finding has been previously reported. The defects in the pericardial cavity in calf No. 2 were similar to the absence of the cavity in a calf with pectoral EC.

Veterinary textbooks routinely define three patterns of arterial branching along the arch of the aorta in the dog, pig, and cow. In cases of bovine cervical EC however, the branching frequently takes on the canine pattern or in some cases a hybrid pattern with features of both the dog and pig patterns.

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Fig. 6. Ventral view, cervical and pectoral region; calf No. 2. Both left and right sternocephalic muscles (Sc) originate from the right cranial end of the sternum. The opening through which the heart protrudes is oval with smooth margins. Bar = 10 cm.

Fig. 7. Dorsal view, sternum, sternal ribs, and pectoral and sternocephalic (Sc) muscles; calf No. 2. The opening is formed cranially by the left pectoral descendens (Pd) and caudally by the cranial border of the sternum. Bar = 5 cm.

Fig. 8. Left view, heart and large arteries; calf No. 2. The branching pattern along the arch of the aorta is comparable to that found in calf No. 1. The fibrous pericardium partially adhered to the epicardium, so that the pericardial cavity was imperfect. L and R = left and right common carotid arteries. Ls and Rs = left and right subclavian arteries. Bar = 5 cm.

However, calves with pectoral EC display either a hybrid pig-cattle branching pattern or the cattle type. In both animals in the present study, a hybrid dog-pig pattern was present (Fig. 9).

The mechanism responsible for cervical EC has been identified as the delayed descent of the heart during embryonic development. In the bovine embryo, descent of the heart is normally completed at about the time a fetus has a crown–rump length (CRL) of 14 mm. In the morphogenetic modification exerted by the embryonic vascular pattern to form the definitive adult pattern, the left subclavian artery switches its site of derivation when the embryo has a CRL of 17–22 mm. This switch involves a change in the site of origin of the left subclavian artery from the aortic arch—a primitive embryonic vascular pattern—to the brachiocephalic trunk. The formation of the bicaudal arteries is initiated after the embryo has a CRL of 25 mm. In bovine fetal life, the CRLs of 14 mm, 17–22 mm, and 25 mm approximately correspond to gestational days 36, 37–39, and 40, respectively. Thus, cervical EC is equated with developmental failure before embryonic day 36, whereas pectoral EC is associated with development subsequent to day 40. In calf Nos. 1 and 2,
Table 1. Defects of heart, artery, veins, and sternum and other anomalies documented in calves with cervical and pectoral ectopia cordis (EC), including the present two cases (Nos. 1, 2).

<table>
<thead>
<tr>
<th>Defects</th>
<th>Cervical EC (8 cases&lt;sup&gt;14&lt;/sup&gt;)</th>
<th>Cervico-pectoral EC</th>
<th>Pectoral EC (one case&lt;sup&gt;16&lt;/sup&gt;)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart position</td>
<td>Cranial to caudal cervical region</td>
<td>Caudal cervical region</td>
<td>Cervico-pectoral region</td>
</tr>
<tr>
<td>Weight (g)</td>
<td>300–580</td>
<td>350</td>
<td>750</td>
</tr>
<tr>
<td>Apex</td>
<td>Double</td>
<td>Double</td>
<td>Single (normal)</td>
</tr>
<tr>
<td>Pericardial cavity</td>
<td>Normal</td>
<td>Normal (expanded)</td>
<td>Imperfect</td>
</tr>
<tr>
<td>Artery branching pattern from</td>
<td>Dog type</td>
<td>Dog-pig type</td>
<td>Dog-pig type</td>
</tr>
<tr>
<td>aortic arch</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Veins</td>
<td>Cranial vena cava</td>
<td>Bilateral</td>
<td>Normal</td>
</tr>
<tr>
<td>Azygos vein</td>
<td>Bilateral, right or left side</td>
<td>Bilateral</td>
<td>Right side</td>
</tr>
<tr>
<td>Pulmonary vein</td>
<td>Several (normal)</td>
<td>Single</td>
<td>Single</td>
</tr>
<tr>
<td>Sternum</td>
<td>Inverted triangle, trapezoid, etc.</td>
<td>Semicircle</td>
<td>Y-shape</td>
</tr>
<tr>
<td>Shape</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Number of sternebrae</td>
<td>13–26</td>
<td>19</td>
<td>10</td>
</tr>
<tr>
<td>Other defects</td>
<td>Torticollis, cleft palate, etc.</td>
<td>Torticollis, cleft palate, and cryptorchidism</td>
<td>Torticollis, cleft palate, and enlarged clitoris</td>
</tr>
</tbody>
</table>

EC is believed to have taken place between fetal days 36 and 40.

In cattle without EC, duplication of the cranial vena cava or vena azygos is an extremely rare anomaly. In duplicative veins have been considered the result of compensation for a morphogenic failure to complete anastomosis between the left and right anterior cardinal and supracardinal veins. As in the calves in this study, a common pulmonary vein was reported, albeit inconsistently, in both calves with cervical EC<sup>3,5</sup> and calves with pectoral EC. As suggested by the fate of embryonic vessels in the modification necessary to establish the basic adult circulatory pattern, the failure of modification at the proper time or the total failure of modification may be the basis for vascular anomalies. In EC, the abnormalities of the vessels near the heart may be attributed to developmental failure in the early embryological stage.<sup>1,2,22</sup>

In a comprehensive treatise on ossification of the bovine skeleton, the early phase of sternal development in the normal bovine fetus was described as follows. At fetal day 50, the precursor of the sternum is identified as bilateral cartilaginous bars. This pair of cartilaginous sternal bars, fully formed by day 53, are fused almost completely by fetal day 64. These observations and the present arterial findings indicate that fusion of the bilateral sternal bars is retarded, if not blocked, by a premature juxtapositioning of the heart at the thoracic inlet. This idea contrasts with an earlier proposition that incomplete fusion of the two halves of the sternum could be regarded as a primary cause of cervical EC.<sup>1,2,13</sup> Because of the strict sequence of events necessary to confer the compromised capacity for differentiation, organogenesis, and fusion of each implicated part in turn, the incompleteness of fusion of the sternal primordial bars is ruled out as playing any important role in the cervical displacement of the heart in bovine EC.

The cause of a broad manubrium and paired sternebrae, widely reported in calves with cervical EC, has been ascribed to the mechanical action of EC itself.<sup>4,11–13,22</sup> The general appearance of the sternums (Fig. 4, B) ranges from that of an inverted triangle to a trapezoid and other shapes. In pectoral EC, however, a ring-shaped sternum forms an opening in the bone (Fig. 4, C), facilitating the protrusion of the heart from the thorax.<sup>1,2,10,16,19,23</sup> In contrast to the sternums characterizing cervical EC and pectoral EC, the sternums of the calves in the present study were disproportionately short for their widths. Furthermore, each of the two sternums had a distinctive shape, one semicircular (Fig. 4, D) and the other Y-shaped (Fig. 4, E), and both resulted from a large incisura on the cranial end. The resultant two sections of each sternum met solely at the xiphoid process or at mid-body. These particular malformations suggest that the manner of attachment...
of the bilateral cervical muscles to the cranial projections of the sternum plays a decisive role in sterno-morphogenesis. In calf Nos. 1 and 2, the EC was classified as intermediate (neither pectoral nor cervical), and the morphologic design of their sternums also reflected the intermediate type of EC (Fig. 4).

To date, only three cases, including the two calves in the current study, of bovine EC intermediate between the cervical and pectoral types have been reported. In the only other case, the main findings were displacement of the heart to a position just cranial to the sternum, left cardiac hypoplasia, double outlet right ventricle, and a partial defect in the pericardium enveloping the heart. EC in calf No. 1 was thought to be closer to the cervical type and that in calf No. 2 closer to the pectoral type than was the other case.

The results of this study support the view that cervical ectopia cordis is determined in the bovine fetus before gestation day 36, that the pectoral type is determined after the 40th fetal day, and that the non-pectoral-noncervical intermediate type occurs between fetal days 36 and 40. Further studies are needed to assess the roles played by breed, genetics, nutritional deficiencies, and detrimental environmental influences on the development of ectopia cordis.

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References


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