

## First report of pentalogy of Cantrell in a calf: a case report

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**ABSTRACT:** This report describes the diagnostic evaluation in a one-week-old, female Simmental twin-calf with the anamnesis of umbilical hernia. Weakness, anaemia, tachycardia and a systolic left sided murmur were significant clinical findings. Based on echocardiography, the animal was diagnosed with pentalogy of Fallot, whereas necropsy revealed the presence of Cantrell's pentalogy with Taussig-Bing syndrome and situs inversus of the liver. Pentalogy of Cantrell is a rare condition described in humans but not yet reported in calves.

**Keywords:** congenital cardiac defect; pentalogy of Cantrell; Taussig-Bing syndrome; situs inversus of the liver; cattle

Pentalogy of Cantrell (PC) is a rare thoraco-abdominal disruption composed of abdominal wall-, diaphragmatic-, pericardial-, sternal-, and heart-defects that was initially described as a specific entity by Cantrell (Cantrell et al., 1958). It is currently classified as an anterior body wall midline developmental anomaly (Carmi and Boughman, 1992) and several variants may occur (e.g. Aslan et al., 2004). To our knowledge no previous report describes this syndrome in cattle, whereas there are a few contributions on Taussig-Bing syndrome in calves (e.g. Hofmann, 1969). Situs inversus of the liver in cattle is mentioned in a single communication (Ries and Koenig, 1988).

### Case presentation

A one-week-old, female Simmental twin-calf was presented to the Clinic for Ruminants at the University of Veterinary Medicine of Vienna with a history of anorexia, abdominal pain and umbilical hernia, which had been surgically closed by the referring veterinarian. The second twin was a fully developed, healthy female calf.

On visual examination, the animal demonstrated exercise intolerance by becoming dyspnoeic follow-

ing mild exertion. The visible mucous membranes were pale. On physical examination, the calf was depressed and in a poor body condition (47 kg body weight). It was febrile (rectal temperature 39.9°C), tachycardic (160 beats/min), and tachypnoeic (120 breaths/min). Auscultation of lung fields and heart revealed increased breath sounds and a grade 5/6 holosystolic murmur with its point of maximal intensity in the pulmonic valve area. On palpation the heart beat could be felt on both sides and caudal to the sternum.

The results of a complete blood count (Cell-analyzer CA 530, Medonic) were within reference ranges besides a mild normocytic and normochromic anemia with a PCV of 0.22 l/l (normal 0.26 to 0.38 l/l) and a haemoglobin concentration of 5.1 mmol/l (normal 5.6 to 8.7 mmol/l). Arterial blood gases (ABL77, Radiometer Copenhagen) revealed a pH of 7.33 (normal, 7.37 to 7.40), hypoxia (pO<sub>2</sub>: 42 mmHg; normal, 70 to 80 mmHg) and hypercapnia (pCO<sub>2</sub>: 52.2 mmHg; normal, 40 to 45 mmHg). Biochemical profiles (Hitachi 911, Hitachi Ltd.; aspartate aminotransferase, gamma glutamyltransferase, glutamate dehydrogenase, bilirubine) were unremarkable apart from a high creatinkinase activity (663 IU/l; normal, <150 IU/l) consistent with muscular damage.

Thoracic radiographs were taken. The cardiac silhouette appeared rounded and globoid. Dorsal positioning of the trachea was evident and an interstitial alveolar lung pattern could be seen. Furthermore, there was some overlap between the cardiac silhouette and the diaphragm. The radiographic findings were summarized as cardiomegaly and mild congestion of the lungs.

Echocardiography (Esaote MyLab30CV, Esaote) revealed an enlarged and hypertrophic right ventricle and atrium, an overriding aorta, a 2 × 2 cm membranous ventricular, and a 3 × 3 cm atrial septal defect (Figure 1). The pulmonary artery seemed narrowed 2 cm distal the valve. Contrast echocardiography was performed with an intravenous injection of 0.9% saline into a jugular vein and resulted in opacification of the right ventricle, followed by simultaneous opacification of aorta, myocardial vessels and pulmonary artery. A negative contrast jet could be seen in the right atrium through the atrial septum. The liver was visible adjacent to the pericardium in the 4<sup>th</sup> and 5<sup>th</sup> intercostal spaces, indicative for diaphragmatic hernia. The cross section of the vena cava caudalis in the 11<sup>th</sup> intercostal space was round with a diameter of 27 mm. Venous congestion was evident. A tentative diagnosis of pentalogy of Fallot was made, and the calf was euthanised.

At necropsy the calf showed a poor nutritional state and several congenital malformations. A complex heart anomaly could be demonstrated: the bulbus aortae was displaced right-laterally and cra-

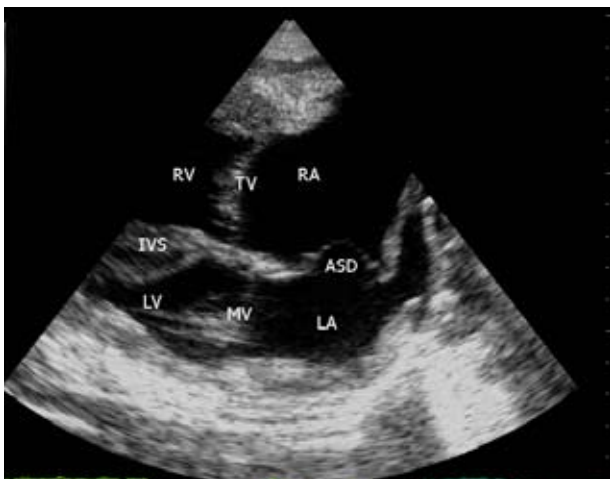


Figure 1. Long-axis echocardiogram of the atrial septal defect (ASD) obtained from the right cardiac window (3.5 MHz). RA = right atrium, TV = tricuspid valve, RV = right ventricle, LA = left atrium, MV = mitral valve, LV = left ventricle, IVS = interventricular septum

nially, and the thick arteria coronaria sinistra arose from the sinus aortae and coursed cranially of the truncus pulmonalis towards the facies auricularis of the heart. The ductus arteriosus (Figure 2) was not obliterated leading blood from the truncus pulmonalis to the aorta. An atrial septum defect with a diameter of about 30 mm bridged by several thin tissue straps was found, and the thickness of the remaining atrial septum did not exceed one mm with exception of its most caudal part.

Both aorta and truncus pulmonalis originated from the right ventricle (double outlet right ventricle). Furthermore, a 15 mm ventricular septum defect situated caudoventrally of the ostium trunci pulmonalis (Figure 2) was detected. The wall of

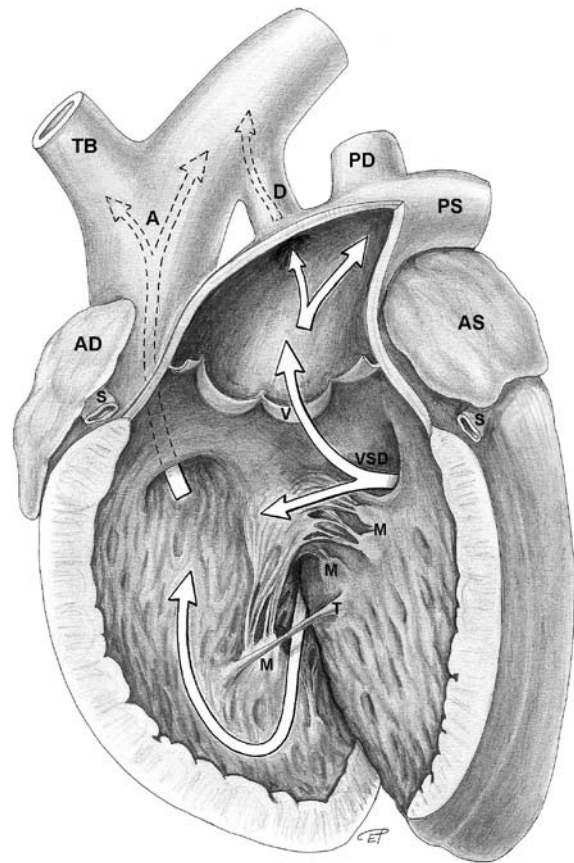


Figure 2. Heart, ventriculus dexter and truncus pulmonalis openend, left-cranial aspect. TB = truncus brachiocephalicus, A = arcus aortae, D = ductus arteriosus, PD = arteria pulmonalis dextra, PS = arteria pulmonalis sinistra, AD = auricula dextra, AS = auricula sinistra, S = arteria coronaria sinistra (cut), V = valvula semilunaris sinistra of valva trunci pulmonalis, VSD = defect in septum interventriculare, M = musculi papillares, T = trabecula septomarginalis, arrows = directions of blood flow

the right ventricle was severely hypertrophic. Blood from the left ventricle must have been directed to the truncus pulmonalis to a great extent, whereas blood from the right ventricle and atrium seemed to have been distributed to both the aorta and the truncus pulmonalis. Histologic evaluation of the myocard showed moderate fibrosis with sporadic scar-formation, moderate endocardial fibrosis and swelling of purkinjefibers with granulated cytoplasm.

The lungs were dystelectatic and hyperaemic with slight alveolar edema and few histiocytes and plasmacells within the alveolar lumina. A crescent shaped ventral diaphragmatic defect with dislocation of the lobus hepatis sinister into the thoracic cavity was present.

The animal's liver demonstrated a mirror-inverted structure (Figure 3) compared to physiological circumstances. It was situated mainly on the left body side, the left lobe projecting into the thoracic cavity through the diaphragmatic defect, and the ligamentum falciforme hepatis was fused with the pericardium. A left and a right umbilical vein were present which connected within the liver (Figure 3). Hepatic histology revealed irregular formation of lobules, incipient cirrhosis with proliferation of fibrotic and bile duct tissues as well as a thickened fibrotic capsule with subcapsular hyperaemia.

On the basis of these results the animal was diagnosed with Cantrell-pentalogy. Taussig-Bing syndrome, a rare congenital deformity with the aorta arising from the right ventricle and pulmonary artery originating from both ventricles, associated with a ventricular septal defect could be demonstrated in this case. The ultrasonographically demonstrated large atrial septal defect could be confirmed at necropsy.

## DISCUSSION

Pentalogy of Cantrell (PC) is a rare congenital syndrome of abdominal wall defect (usually omphalocele), lower sternal defect, diaphragmatic pericardial defect, anterior diaphragmatic defect, and intracardiac abnormalities. First described by Cantrell in 1958, the syndrome occurs sporadically, with variable degrees of expression (Cantrell et al., 1958). The proposed pathogenesis involves a defect in embryogenesis between 14 and 18 days after conception, when the splanchnic and somatic mesoderm are dividing. Many associated anomalies have been reported in fetuses with PC, including cranial and facial anomalies, clubfeet, malrotation of the colon, hydrocephalus, and anencephaly.

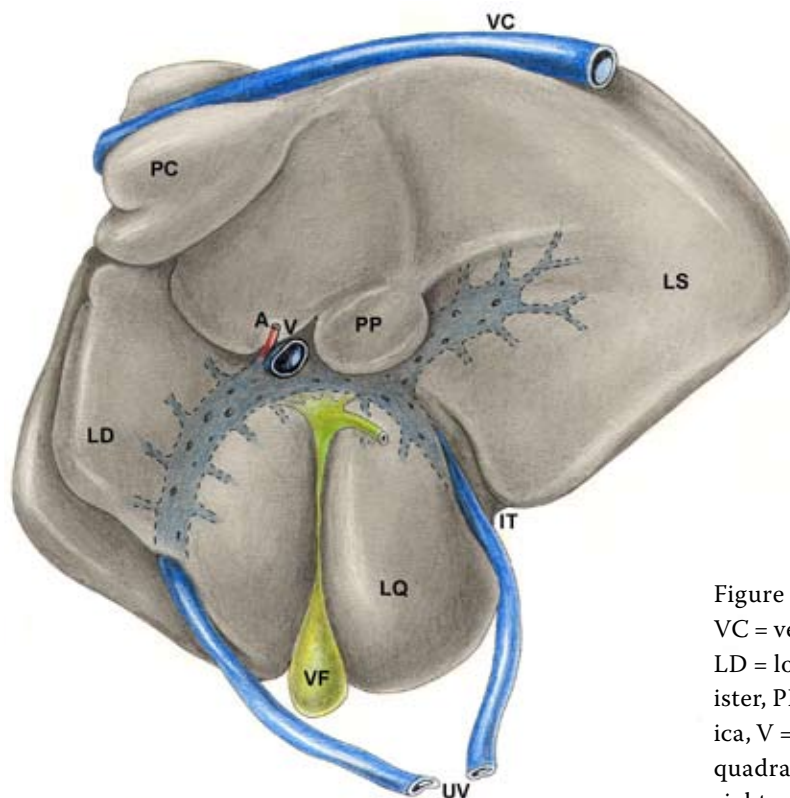


Figure 3. Liver, facies visceralis, right = cranial. VC = vena cava caudalis, PC = processus caudatus, LD = lobus hepatis dexter, LS = lobus hepatis sinister, PP = processus papillaris, A = arteria hepatica, V = vena portae, VF = vesica fellea, LQ = lobus quadratus, IT = incisura ligamenti teretis, UV = right and left umbilical vein

Chromosomal abnormalities have also been associated with the syndrome (Carmi and Boughman, 1992). Less than 90 cases have been reported in the literature in humans, and even fewer have had the complete syndrome confirmed. Only four of the 90 cases involved twins and only two of them described discordance for the anomaly (Rashid and Muraskas, 2007) like in the present report.

Diagnosis of the complete syndrome requires the five criteria described by Cantrell, but incomplete variant forms exhibiting three or four of the features have been reported. The sternal defect can range from absence of the xiphoid to cleaving, shortening, or absence of the entire sternum. The abdominal defect can range from a wide rectus muscle diastasis to a large omphalocele. The most common intracardiac defects are atrial septal defect, ventricular septal defect, and tetralogy of Fallot. Differential diagnosis include isolated ectopia cordis and isolated abdominal wall defects. The prognosis appears dismal, but may be related to the extent of the ventral wall, sternal, and cardiac defects (Bryke and Breg, 1990).

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Received: 2008–09–17

Accepted: 2008–12–11

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