Case report: Persistent truncus arteriosus with intact ventricular septum and other cardiovascular malformations in a calf

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SUMMARY
A 3-day old, male Holstein calf presented for necropsy with a history of respiratory distress, recumbency and failure to suckle since birth. Postmortem examination revealed the absence of an aorta and a single artery arising from the right ventricle that became the pulmonary trunk, which branched into the main pulmonary artery and rose to two pulmonary arteries (type-1 persistent truncus arteriosus). Other related findings included intact ventricular septum, left ventricular hypoplasia, left coronary artery originating from brachiocephalic trunk, absence of right coronary artery and patent foramen ovale. Microscopically, there was moderate, diffuse subendocardial fibrosis, myocardial degeneration, necrosis and multifocal mononuclear cell infiltrations, which were most prominent in the left ventricle wall. Additionally there was marked medial thickening of arterial and arteriolar walls in the lungs. Pulmonary parenchymal changes included moderate edema, vascular and interstitial congestion, hemosiderin laden macrophages and diffuse suppurative pneumonia. Consequently, the concurrence of type-1 persistent truncus arteriosus, left ventricular hypoplasia, single right coronary artery and anomalous origin of the right coronary artery was unusual in many respects and contributed to this calf’s failure to thrive and early death.

Keywords: bovine, left ventricular hypoplasia, persistent truncus arteriosus

Introduction
Congenital defects are the abnormalities present at birth resulting from development. They can affect all or parts of several system or isolated organ [22]. Persistent truncus arteriosus (PTA), otherwise known as truncus arteriosus communis or common aortico-pulmonary trunk, is an uncommon congenital cardiac malformation describing the absence or complete failure of the normally divided embryonic truncus arteriosus (TA) into aortic and pulmonary trunk. A single truncus originates from the left, right ventricle or interventricular septum and supplies systemic, coronary and pulmonary blood flow [7, 22]. The incidence of cardiac malformations in bovine fetus is reported as 0.8% [19].

In humans, the incidence of cardiovascular malformations is 0.62% of live births and PTA consisting of approximately 1.33% of total cardiovascular malformations [24]. As PTA has been reported sporadically in veterinary literature, its true incidence is unknown in animals [10, 13, 21]. Up till date, PTA has been documented in 9 calves [5, 14, 19, 24-26], lambs [13, 20], foals [9, 10, 17], dogs and cats [3, 6, 7, 21, 27], a pig [16], a monkey [2] and a baboon [12].

To date, PTA has always been reported concurrently with ventricular septal defect in animal species. Other associated reported malformations and lesions are patent foramen ovale [23, 26], atrial septal defect [16], main pulmonary artery hypoplasia [17], diprosopus [5], dissecting aneurysm and cardiac tamponade [13] and ventricular aneurysm [23].

This paper describes concurrent occurrence of PTA with intact interventricular septum, left ventricular hypoplasia, patent foramen ovale, atrioventricular fistula and coronary artery malformations in a male Holstein calf.

Case report
A 3-day-old, male Holstein calf was reported to be showing respiratory distress and recumbency with no
sucking reflex since the birth and colostrum was given with the aid of feeding bottle. Referring veterinarian injected intramuscular antibiotic (Baytril® 100, 100 mg enrofloxacin in 1 mL, Bayer) injection for 2 previous days with diagnosis of pneumonia. After death on third day, he was presented for the necropsy to the Pathology Department of Veterinary Medical School, Firat University. He weighed 23.80 kg, having a poor body condition, and was the third offspring of a naturally inseminated dam. The previous calves of the dam had no clinical problem.

Results

Postmortem examination showed that the calf was underdeveloped and the muzzle was markedly cyanotic. The heart was remarkably enlarged with globoid appearance weighing 285 gr. Its apex was constituted completely by the right ventricle. There was no aorta arising from the left ventricle. The left ventricle was extremely hypoplastic measuring 2.51x1.43 cm and weighing 40 gr. The left ventricular wall was considerably thickened and showed concentric hypertrophy. The weight of interventricular septum was 85 gr. The vertical and lateral thickness of the left ventricular wall was 3.6 cm and 2.5 cm, respectively. There was a fistular connection measuring 5 mm in diameter in the lateral margin of left ventricle communicating with the left atrium at the level of pulmonary vein. The right ventricle was dilated by 6.0x5.8 cm, weighing 155 gr and completely formed the apex. The foramen ovale between the atria was patent having a flap (1.92x1.0 cm) and fenestrated hole (6.0x5.0mm) in size (Figure 1). TA exhibited tricuspid valves. Single large arterial vessel having 2.8 cm diameter originating from the right ventricle became pulmonary trunk at 7.30 distance cm and branched into main pulmonary artery and then giving rise to two pulmonary arteries. After that, the arterial truncus at 12.30 cm distance from the origin gave the branch resembling ascending aorta whose first division was brachiocephalic (arterial) trunk (Figure 2). There was apparently no right coronary artery. The area was vascularized by the ramifications of ramus flexus of the left coronary artery. The first thick division of brachiocephalic trunk directing to the right ventricle gave rise the left coronary arterial ramus, in 34 mm in length, and ended at the level of interventricular sulcus (Figure 3). According to criteria used in humans [8], the malformation was classified as Type-1 PT. Both of the atria and the right ventricle were moderately dilated. The endocardium of left ventricle was moderately thickened and opaque. The lungs were heavy and showed severe, diffuse hyperemia, edema and hard consistency. The liver was also hyperemic in appearance. The abomasum and intestines were completely full of colostral ingesta. For microscopic examination, tissue samples from the TA, ascendent aorta, liver, lungs, pancreas, brain, small-large intestines and kidneys were fixed in 10% formalin and processed routinely. The sections were stained with hematoxylin and eosin (H&E), and selected heart sections were also stained with Masson's trichrome.
Microscopic examination in the heart consisted of myofibrillary degeneration, focal necrosis and focal post-necrotic mononuclear cell infiltrations in both ventricles. There was severe to moderate subendocardial fibrosis in the left ventricle (Figure 4) and marked thickening in the vessel walls and narrowing of the lumen in intramural coronary arteries and mild to moderate perivascular fibrosis (Figure 5), whereas in the right ventricle there was mild perivascular edema and fibrosis.

The changes in the lung included marked medial thickening of the wall of arteries and arterioles, perivascular edema and mild fibrosis. In alveoli, there was moderate, multifocal accumulation of neutrophils and lymphocytes, hyperemia, hemosiderin laden macrophages, edema and congestion. The liver also showed severe to moderate, diffuse hyperemia.

**Discussion**

To our knowledge, this report represents the first case of PTA with intact interventricular septum and left ventricular hypoplasia in animals. However, concurrence of these malformations was reported as a rare variant in a few human infants [1]. In these reports; TA arising from left ventricle or right ventricle [1] co-existed with the opposite ventricular hypoplasia. Left ventricular hypoplasia defines a congenital and lethal heart abnormality occurring usually association with obstruction of left ventricular outflow [15]. Taken together; our findings support the conclusion that left ventricular hypoplasia seems to be secondary to obstruction of left ventricular flow. PTA is divided into 4 anatomic subtypes according to location of pulmonary artery and TA in humans [8]. The present case resembled Type-1 PTA of human classification due to a single pulmonary trunk dividing into 2 pulmonary arteries and a single ascending aorta.

The formation of left atroioventricular fistula and patent foramen ovale in the present report might be related with high intracardiac pressure in left heart.

Genetic factors and familial occurrence have been reported in keeshond dogs. A genetic study suggested that specific mutations in oligenic genes including CFA2, CFA9 and CFA15 may be responsible for conotruncal heart malformations. These mutations might be spontaneous or induced by teratogens [27]. The clinical signs in PTA are directly related with the proportion of pulmonary to systemic blood flow [7]. In the present case, due to intact ventricular septum, there is insufficient blood flow to the both pulmonary artery and aorta. Macroscopic lesions including cyanosis, left to right shunting and pulmonary congestion-edema were consistent with heart failure. Blood flowing from right ventricle through TA enter either pulmonary circulation or systemic circulation. Incase of pulmonary
vascular resistance is not high, blood flows into pulmonary circulation, and heart failure ensues and resulting in death in a few days. When pulmonary hypertension is present, desaturated blood from the right ventricle is ejected into systemic circulation, resulting from hypoxemia and cyanotic membranes [7]. In the present report, pulmonary circulatory inadequancy and pulmonary hypertension were marked by cyanosis and thickening of arterial wall and severe purulent pneumonia. The occurrence of pneumonia in present and previous studies might be explained as a complication of heart failure and pulmonary edema.

In humans; 42% of TA originates from the right ventricle, 42% of TA from left ventricle and 16% originates from interventricular septum [4]. Similar to presented case, TA originated from the right ventricle is predominantly reported form of PTA in animals [6, 7, 9, 13, 16, 17, 21, 25, 26].

The PTA is usually fetal in newborn period in animals. In humans; 80% or more infants die within first year of life without surgical intervention [24]. In animals, there are three published PTA cases having long term survival including a dog with Type-3 (8 years), in a cat with Type-1 (6 year) and in a steer with Type-2 (2 years) [14, 21]. The common malformation in all of these cases is a large ventricular defect.

Currently, pathogenesis of PTA is not fully understood, but appears to involve the inhibition of the migration of cardiac crest cells resulting in failure of septation of TA [22, 27].

The coronary artery arising from brachiocephalic artery and absence of right coronary artery has never been reported earlier either alone or simultaneously with PTA in animals. However, a single coronary artery (like the present case) arising from posterior aspects of truncus pulmonalis was noted among the 4 of 30 human PTA cases, corresponding 13% of total cases [4]. As there is possibility of neural crest involvement in coronary arterial systemdevelopment [18] hence, it is also reasonable to assume that coronary artery malformations could also arise as the same mechanism with PTA.

In conclusion, the concurrence of type-1 PTA, left ventricular hypoplasia, single right coronary artery and anomalous origin of the right coronary artery was unusual in many respects and combination of these factors along with pneumonia might contribute to the early prenatal death in the calf.

References


