

## TRUNCUS ARTERIOSUS PERSISTENS IN AN ARABIAN FOAL

### *Truncus arteriosus persistens bij een Arabisch volbloed veulen*

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#### ABSTRACT

**A ten-day-old Arabian filly was examined because of signs of weakness, respiratory distress and cyanosis. On auscultation a grade V/VI holosystolic murmur was found on both sides of the thorax. The point of maximal intensity was at the right cranial part of the thorax. On the basis of clinical and echocardiographic examination, a differential diagnosis of truncus arteriosus, pseudotruncus arteriosus, tetralogy of Fallot or aorticopulmonary septal defect was made. Despite the poor prognosis, the owner refused euthanasia. The condition of the foal deteriorated progressively and the foal died at two months of age. Necropsy demonstrated a persistent truncus arteriosus. This paper also describes the characteristic echocardiographic differences which can be observed between a normal heart and a heart with a truncus arteriosus.**

#### SAMENVATTING

Een tien dagen oud Arabisch volbloed veulen werd aangeboden wegens de klacht van zwakte, dyspnoe en cyanotische mucosae. Tijdens de auscultatie werd een luid holosystolisch bijgeruis opgemerkt ter hoogte van beide zijden van de thorax. Het bijgeruis was het duidelijkst hoorbaar ter hoogte van het craniale deel van de rechterthoraxwand. Aan de hand van het klinisch onderzoek en de echocardiografische bevindingen werd een differentiaaldiagnose van truncus arteriosus, pseudotruncus arteriosus, tetralogie van Fallot of een aorticopulmonair septumdefect gesteld. Ondanks de slechte prognose besloot de eigenaar het veulen aan te houden, maar de toestand verslechterde progressief en het veulen stierf op de leeftijd van twee maanden. Tijdens de autopsie werd de aanwezigheid van een truncus arteriosus vastgesteld. In dit artikel worden naast de algemene beschrijving van een veulen met een truncus arteriosus ook voornamelijk de specifieke echocardiografische bevindingen vergeleken van een normaal hart met die van een hart met een truncus arteriosus.

#### INTRODUCTION

In horses, the prevalence of congenital heart anomalies is rather low and until now no underlying causes have been established. In human medicine, multifactorial disorders such as the combined effect of a genetic predisposition, gene mutations or other factors such as maternal infection, age of the mother, nutritional status, hypoxia or trauma are believed to play a major role (Olson and Strivastava, 1996; Ya *et al.*, 1997; Reef and McGuirk, 2002). Compared to ventricular septum defect, persistent truncus arteriosus is a relatively rare anomaly in foals (Lombard *et al.*, 1983; Reef, 1985). A persistent truncus arteriosus is characterized by one big vessel that gives rise to the

outflow tract of the left and right ventricles. In foals, the antemortem diagnosis of a persistent truncus arteriosus has already been described in Arabian foals (Bayly *et al.*, 1982; Sojka, 1987), a Warmblood foal (Tschudi *et al.*, 1997), a Quarter Horse foal (Steyn *et al.*, 1989) and a Bashkir Curly foal (Stephen *et al.*, 2000). In other animals, such as a calf (Schwarzwald *et al.*, 2003) and a cat (Nicolle *et al.*, 2005), ante mortem diagnosis of a persistent truncus arteriosus has been described occasionally. Post-mortem, this anomaly has been described in dogs (Chen *et al.*, 1972, van Mierop *et al.*, 1978), cats (Buergelt and Suter, 1968, van de Linde-Sipman *et al.*, 1973), a lamb (Milstein *et al.*, 1982), calves (Heath and Kruketti, 1979; Kemler and Martin, 1972; Reppas *et al.*, 1996;

Sandusky and Smith, 1981; West, 1988), foals (Greene *et al.*, 1975), piglets (van de Linde-Sipman and Wensing, 1972), and a Rhesus Monkey (Brandt *et al.*, 2002).

This paper describes clinical, echocardiographic and post-mortem findings of a persistent truncus arteriosus in an Arabian foal.

CASE HISTORY AND CLINICAL EXAMINATION

An Arabian filly was born at full-term gestation. Although the foal suckled normally within 1 hour after birth, weakness and shortness of breath were noticed. At presentation, the foal was in normal body condition (48 kg). Physical examination revealed a body temperature of 38.3°C, cyanotic mucous membranes and a fast respiratory rate (40 breaths/min) with enforced respiration sounds. On auscultation a regular heart rate of 100 beats/min and a loud (grade V/VI) holosystolic murmur was found on both sides of the chest with the point of maximal intensity over the cranial right thorax. A thrill was palpable on both sides of the thorax. Peripheral pulses were weak. Hematological and biochemical parameters were within normal references, except for a low serum immunoglobulin G concentration (4 g/l).

Echocardiographic examination (GE Vingmed, England, 2.5 MHz) was performed on the right side of the thorax but it was complicated by significant lung interference and the distress of the foal. At the fourth intercostal space, a four chamber view indicated a small left atrium (Figure 1). On the left ventricular outflow tract view, a large vessel (TA) which was assumed to be the aorta could be observed (Figure 2a). On the right parasternal oblique view obtained from the right fourth intercostal space with the transducer pointing dorsally, a connection was observed between the root of the large vessel (TA)

and the right ventricle (Figure 3a). This finding was confirmed by Color-Flow Doppler.

Because of the foal’s agitation, the echocardiographic examination was terminated prematurely. A presumptive diagnosis of a severe congenital heart anomaly such as truncus arteriosus, pseudotruncus arteriosus, tetralogie of Fallot or aortic-pulmonary septal defect (Valdes-Martinez *et al.*, 2006) was made on the basis of the clinical examination and this limited echocardiographic examination. Despite the grave prognosis, the owners decided to take the foal back home and to follow its evolution. No medical treatment was given. The foal’s condition deteriorated and it died at the age of 2 months.

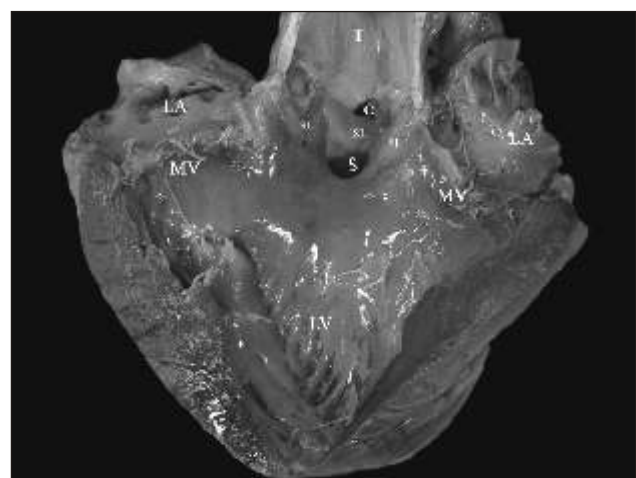


Figure 1. The heart of a ten-day-old Arabian filly with a truncus arteriosus communis (T). A centrally located tun-cus arteriosus communis, 4 cm in diameter and with 3 large semilunar valves (SL), was found arising from both ven-tricles at the heart base. A 2 cm in diameter ventricular septum defect (S) was present immediately below the SL valves. The SL valve showed no macroscopic anomalies. LA: left atrium. LV: left ventricle. MV: mitral valve. C: co-ronary artery.

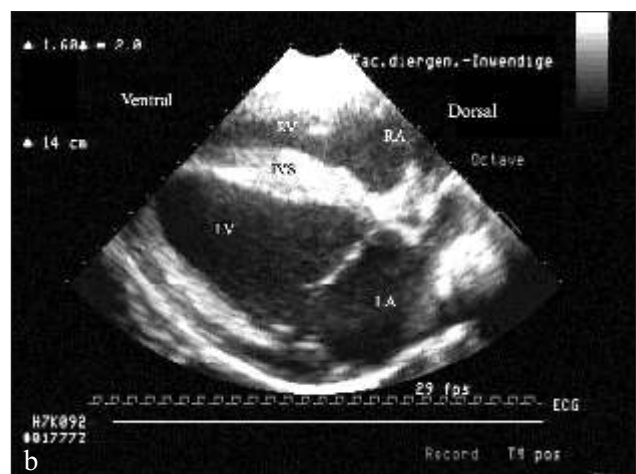
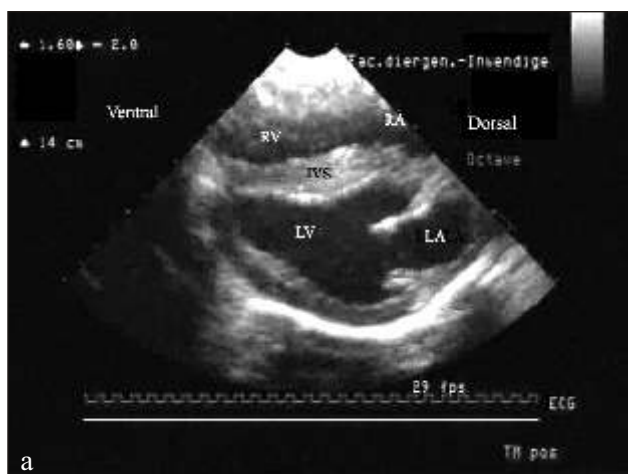
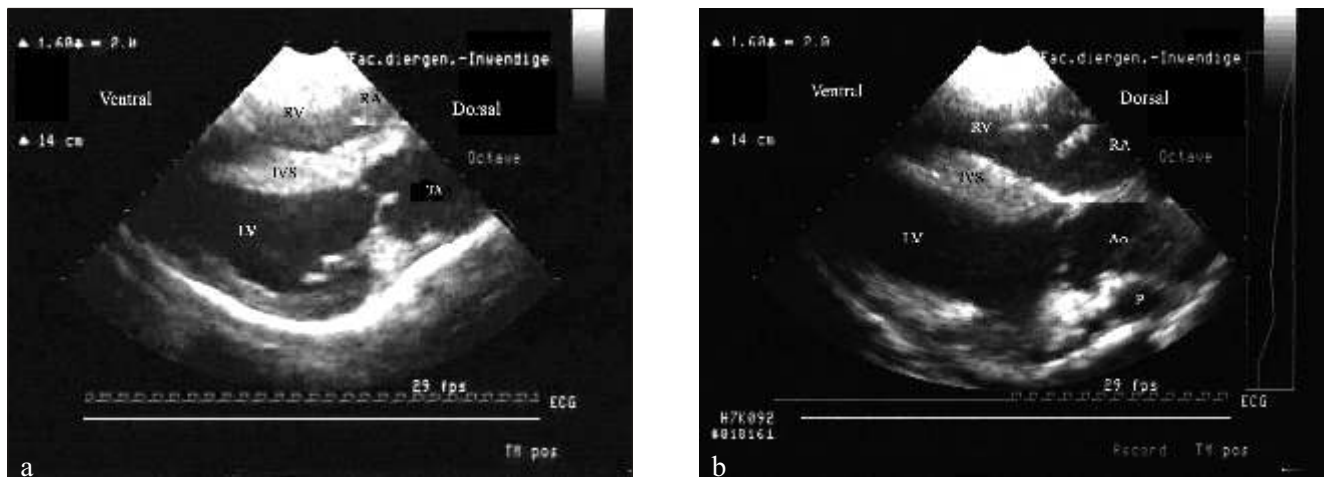


Figure 2. A four-chamber view of a 10-day-old foal with a truncus arteriosus persistens (a) compared to the same view in a normal 10-day old foal (b). This echocardiogram was obtained with a 2.5-MHz sector-scanner transducer at a displayed depth of 14 cm. RA: right atrium. RV: right ventricle. LA: left atrium. LV: left ventricle. IVS: interventricular septum.



**Figure 3.** Right parasternal long-axis view of the left ventricular outflow tract of a 10-day-old foal with a truncus arteriosus (a) compared to the same view in a normal 10-day old foal (b). This echocardiogram was obtained with a 2.5-MHz sector-scanner transducer at a displayed depth of 14 cm. No truncus pulmonalis could be visualized in the view of the foal with the truncus arteriosus persistens (a). The right side of the echocardiogram is dorsal and the left side is ventral. RA: right atrium. RV: right ventricle. LV: left ventricle. Ao: aorta. IVS: interventricular septum. TA: truncus arteriosus persistens. P: truncus pulmonalis.

#### POSTMORTEM FINDINGS

The heart and coronary vessels were enlarged. A centrally located truncus arteriosus communis, 4 cm in diameter and with 3 large semilunary valves (SL), was found arising from both ventricles at the heart base. A 2 cm in diameter ventricular septum defect was present immediately below the SL valves. The SL valve showed no macroscopic anomalies (Figure 1). At five cm from the SL valves a vessel, 1.3 cm in diameter, was found to originate from the truncus arteriosus. This vessel was 2 cm long before it bifurcated to supply both lungs.

#### RETROSPECTIVE VIDEO ANALYSIS

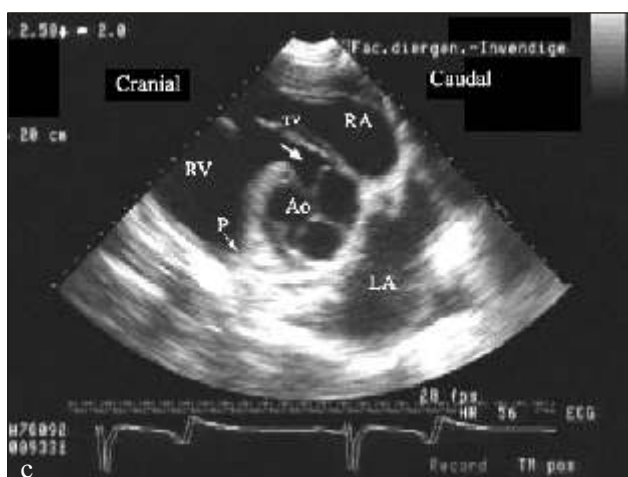
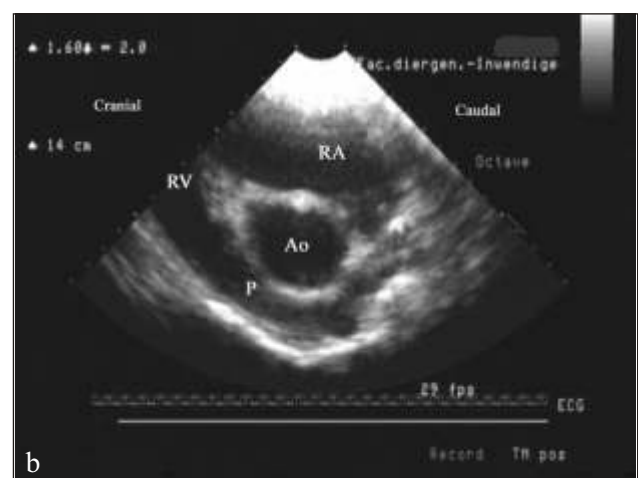
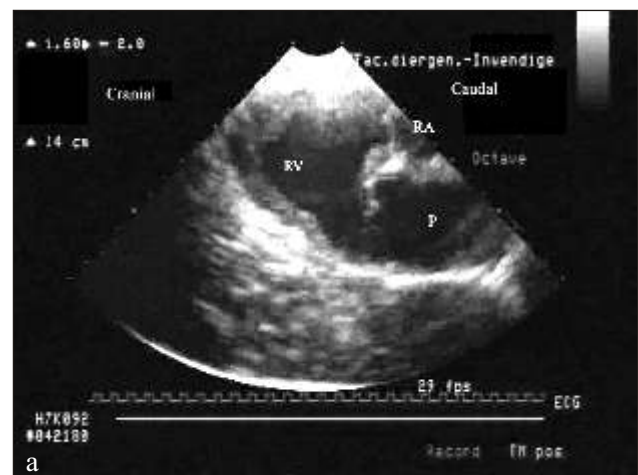
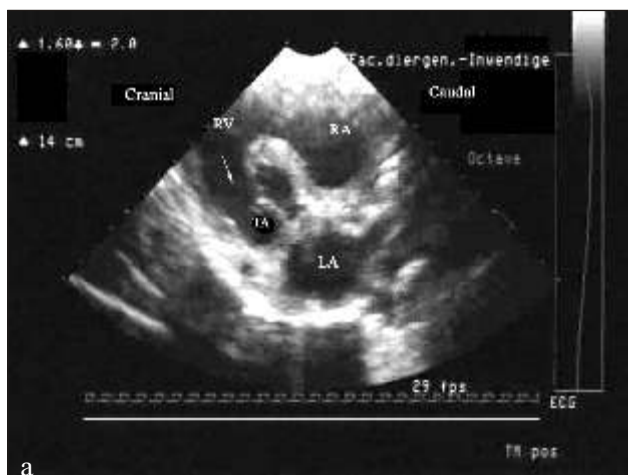
On the basis of the post-mortem results, a retrospective video analysis of the echocardiographic examination was performed. On a standard four chamber view, the left atrium is smaller compared to that of a normal 10-day-old foal (4.65 cm vs. 6.56 cm, measured during maximal ventricular diastole) (Figure 2a vs. Figure 2b).

One large vessel (TA) appears at the normal location of the aorta on the right parasternal long-axis view of the left ventricular outflow tract (Figure 3a vs. Figure 3b) and on the right parasternal oblique view from the right fourth intercostal space with the transducer pointing dorsally (Figure 4a vs. Figure 4b). When compared to the normal aorta, this vessel is larger than normal (4.03 cm versus 3.23 cm in diastole; 4.32 cm versus 3.50 cm in systole, measured in a normal 10-day-old foal). In addition, on the right parasternal long-axis view of the left ventricular outflow tract (Figure 3a vs. Figure 3b) and on the right ventricular inflow-outflow view (Figure 5a vs. Figure

5b), no pulmonary artery could be visualized. The right parasternal oblique echocardiogram obtained from the right fourth intercostal space with the transducer pointing dorsally shows that this vessel was connected to the right ventricle (Figure 4a vs. Figure 4b). However, this opening was located more ventrally compared to the view of a 2-month-old foal with a membranous ventricular septum defect (Figure 4a vs. Figure 4c)

#### DISCUSSION

At 20 days of gestation the mammalian embryo is about 5 to 6 mm in length and at this stage the physicochemical processes cannot adequately provide for the enormously increasing metabolic needs of the embryo. Therefore a functional circulatory system needs to develop. The heart has to grow from a single tube into a complex four chambered organ, possessing four sets of valves. In brief, the single tube differentiates first into endocardium, myocardium and pericardium, and subsequently divides into a left and a right atrial part and a bulboventricular part. The embryo is then about 23 days old and the preliminary heart starts to beat. In the next step the real structural formation of the different compartments of the heart begins to develop. The cranial third of the bulboventricular part will form the aortic sac, the caudal third is the early embryonic left ventricle and the mid-portion will develop into the bulbus cordis. Because the heart tube is fixed at both ends and the tube grows rapidly, it is forced to bend and split up in a pre-programmed manner to adapt itself to the available space. The proximal part of the bulbus cordis becomes the right ventricle, the mid-third portion forms the outflow portion of both ventricles

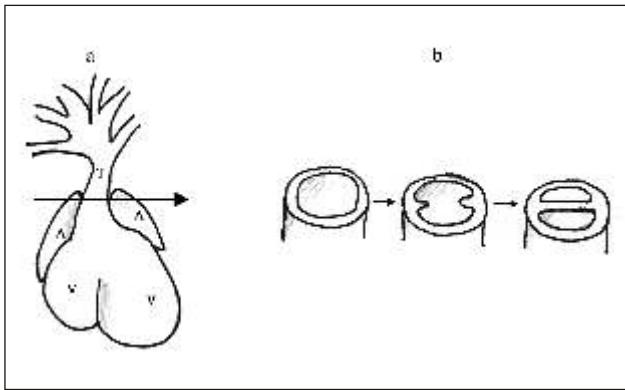


**Figure 4.** A right parasternal oblique echocardiogram obtained from the right fourth intercostal space with the transducer pointing dorsally at the level of the aortic valves of a 10-day-old foal with a truncus arteriosus persistens (a) compared with the same view in a normal 10-day old foal (b) and compared to the same view of a 2-month old foal with a membranous ventricular septum defect (c). The (a and b) echocardiograms were obtained with a 2.5 MHz sector scanner transducer at a displayed depth of 14 cm. The echocardiogram (c) was obtained with a 2.5 MHz sector transducer at a displayed depth of 20 cm. On Figure 4a a connection is observed between the aortic root and the right ventricle (arrow). However, compared to Figure 4c, the connection (arrow) is located more ventrally. RA: right atrium. TV: tricuspid valve RV: right ventricle. Ao: aorta. LA: left atrium. P→: truncus pulmonalis.

**Figure 5.** The right ventricular inflow-outflow echocardiogram of a 10-day-old foal with truncus arteriosus persistens (a) compared to the same view of a normal 10-day old foal (b). This echocardiogram was obtained with a 2.5-MHz sector-scanner transducer at a displayed depth of 14 cm. On Figure 5a, no aorta is present. RA: right atrium. RV: right ventricle. Ao: aorta TA: truncus arteriosus P: pulmonary artery.

and the terminal third develops into the proximal parts of the ascending aorta and the pulmonary trunk. The aortopulmonary septum (Figure 6a and b) is formed by migration of some cells from the cranial neural crest to the 3<sup>th</sup>, 4<sup>th</sup> and 6<sup>th</sup> pharyngeal arches. At the same time, some of the cells of the same region migrate in the direction of the pharyngeal apparatus, including thymus, parathyroid and thyroid glands. However, in the event of persistent truncus arteriosus, the strict strategies of septation fail at the stage when the embryo is about 8.5 to 9.5 mm and approximately 25 to 28 days old (Netter, 1969; Kirby *et al.*, 1985; Kirby and Waldo, 1990; Waldo *et al.*, 1998; McGeady *et al.*, 2006).

Because only one common trunk and an interventricular septum defect is present, a loud holosystolic murmur at both sides of the chest is audible. With this anomaly, a mixture of oxygenated blood and deoxygenated blood flows to the pulmonary trunk supplying both lungs, as



**Figure 6. Ventral view of the embryonic heart (a) and schematic representation of the formation of the aortico-pulmonary septum (b) (embryological stage between 25 and 28 days). T: truncus arteriosus. A: atrium cordis. V: ventriculus cordis.**

well as to the general circulation. Lungs receive hypertensive blood, which can induce lung edema. Therefore patients can go into respiratory distress and mucus membranes can become cyanotic.

Correct formation and function of the heart requires a multitude of specific cell and tissue interactions, and so each region of the tube is programmed to become a particular part of the heart. In some rare cases, however, any divergence of these specific interactions results in a congenital malformation of the heart. Only 0.5-1% of these abnormal embryos survive, and 10% of those that survive are eliminated by stillbirths and up to 20% die and abort spontaneously (Hoffman and Christianson, 1978; Hoffman, 1995). In a study of 8954 necropsies of foals that died as fetuses or that were euthanized as foals or yearlings, 3.5 % had different forms of congenital cardiac malformations, which represents an incidence of 0.23 % (Crowe and Swerczek, 1985).

In a newborn foal it is always indicated to make a complete physical examination. Many clinical parameters are similar in the adult and neonatal animals, but special attention should be paid to identification of any congenital malformation. By auscultation of the neonatal heart it is normal to hear a machinery murmur of a patent ductus arteriosus until an age of 4 days. After this age, even in the absence of any detectable cardiac disease, foals may present a soft systolic murmur over the left heart that may persist for up to 60 days (Lombard, 1990). However, a complete echocardiographic examination should be performed when a loud murmur is found.

In cases of common truncus arteriosus persistens, echocardiography reveals a single large vessel originating from the right ventricle above the interventricular septum and a large defect at the top of the interventricular septum can be visualized (Bayly *et al.*, 1982; Schwarzwald *et al.*, 2003; Sojka, 1987; Stephen *et al.*, 2000; Steyn *et al.*, 1989; Tschudi *et al.*, 1997, Nicolle *et al.*,

2005). Echocardiographic examination in this foal was difficult to perform and had to be aborted because of agitation and increasing respiratory distress. Therefore, only a presumptive diagnosis of a severe congenital heart anomaly could be made.

The retrospective video analysis revealed additional information about the anomaly. The opening between the aortic root and the right ventricle could only be observed on a right parasternal oblique echocardiogram from the fourth intercostal space with the transducer pointing dorsally. With the presence of persistent truncus arteriosus, it was obvious that a normal pulmonary artery inflow could not be visualized. The presence of a smaller left atrial diameter can be explained because of a decreased blood flow towards the lungs and therefore a decreased blood flow returning from the lungs to the left atrium.

In human medicine, a classification is made based on the anatomical origin of the pulmonary vessels from the common trunk. The oldest classification dates from 1949 and is called the Collett and Edwards Classification (Collett and Edwards, 1949), but an adapted classification has been described by Van Praagh and Van Praagh (1965). The Collett and Edwards Classification describes four different types based upon the origin of the pulmonary artery. In Type I, a short pulmonary trunk emerging from the truncus arteriosus persistens gives rise to both pulmonary arteries. In Type II, the right and left pulmonary arteries arise posteriorly and individually from the truncus, but close to each other. In Type III, the orifices of the pulmonary arteries arise more laterally and individually from the truncus, but widely separated from each other. Type IV is considered to represent a form of pulmonary atresia with ventricular septal defect, also called Tetralogy of Fallot. The Van Praagh and Van Praagh Classification Type A1 is the same as the Collett and Edwards Type I. In Type A2, the right and left pulmonary arteries arise separately from the truncus (as Type II and III in the Collett and Edwards Classification). In Type A3, a single pulmonary artery arises from the truncus while the opposite lung is supplied by collaterals, and in Type A4 there is a hypoplastic aortic arc with a large patent ductus arteriosus arising from the truncus and supplying the descending aorta. In the currently described foal, a short pulmonary trunk gives rise to both pulmonary arteries and is therefore classified as a Type I or Type IA. In human literature, the prevalence of Type I is the highest compared to the other types, and represents 47% to 77% of all cases (Butto *et al.*, 1986; Houston *et al.*, 1981; Van Praagh and Van Praagh, 1965).

In conclusion, echocardiographic examination is a very important tool for diagnosing the most common neonatal anomalies. However, examination of a neonatal patient in distress can be challenging. Exact anatomical

knowledge of the normal cardiac structures and congenital anomalies is primordial for recognizing the exact nature of the malformation.

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