Cor biloculare with truncus arteriosus communis: autopsy case report with literature review

Dorota Sopková1, Silvia Farkašová İannaccone1,*, Daniel Farkaš2

Abstract: Cor biloculare associated with a persistent truncus arteriosus is a very rare congenital heart disease. Referred to also as a two-chambered heart, cor biloculare is a malformation characterized by the presence of a single atrium and single ventricle communicating through a common atrioventricular valve. In association with truncus arteriosus communis, instead of separate aorta and pulmonary artery originating from the common ventricle, there is a single arterial trunk present, giving rise to coronary, pulmonary and systemic arteries. This combined malformation belongs to the group of cyanotic congenital cardiac diseases with a death of the affected individual occurring usually in the neonatal period. We present the autopsy findings of a premature female infant with a cor biloculare associated with truncus arteriosus communis born at a gestational age of 23 weeks, who died a day and a half after the birth.

Key Words: cor biloculare, truncus arteriosus communis, premature infant, autopsy findings.

INTRODUCTION

Cor biloculare with a persistent truncus arteriosus is a rare cyanotic congenital cardiac disease. Two-chambered heart is characterized by a single atrium communicating through a common atrioventricular (AV) valve with a single ventricle. Instead of ascending aorta and pulmonary artery a single arterial trunk is originating in the common ventricle, then continuing to form an aortic arch and descending aorta [1]. Cor biloculare is commonly associated with a presence of other malformations, such as dextrocardia with or without transposition of abdominal organs, asplenia, or pulmonary and systemic venous anomalies [2]. The authors present the autopsy findings in a premature female newborn infant with cor biloculare associated with truncus arteriosus communis, who died shortly after the birth.

CASE REPORT

Premature female infant was born at a gestational age of 23 weeks by a spontaneous vaginal delivery. Apgar scores upon birth were 2 and 4, the birth weight was 440 g and length 30 cm. Severe hypothermia (30°C) was present despite the warming efforts. Echocardiography proved congenital heart disease, which was suspected to be the atrioventricular septal defect. Based on the contrast radiography of the upper gastrointestinal tract tracheal atresia with tracheoesophageal fistula was also suspected. The course of the treatment was very short, and due to extreme prematurity, severe respiratory and circulatory failure, and the presence of congenital defects the prognosis was assessed as infaust. With further attempts of invasive treatment considered as futile, and based on the informed consent of the parents, only the palliative care was provided. The infant died approximately 34 hours after the birth.
At autopsy the body was that of a premature female newborn infant. Her length (30 cm) and weight (436 g) were within the normal range for the stated gestational age of 23 weeks. Infant size was determined to be normal according to the measurement of head circumference (20 cm), as well as the abdominal (16 cm) and thoracic circumference (17 cm). No sign of a disproportionate growth of extremities was observed. Skin was fragile and with no appendages. External examination did not determine any congenital or developmental abnormalities.

Upon opening the thoracic cavity the heart, enclosed in the pericardial sac, was lying in the centre of the chest. The pericardium didn’t contain any fluid. The heart itself weighted 2 g (Fig. 1). Only one atrial appendage was present, attached to the surface on the right side of the heart. Upon opening the heart there was one atrium and one ventricle present with no traces of interatrial septum, nor interventricular septum. The wall thickness of the common ventricle was 0,2 mm (Figs 2, 3). Common atrium and common ventricle were communicating through a single valve composed of three cusps, with each cusp attached separately to the three papillary muscles situated in the common ventricle. From the common atrium originated superior and inferior vena cava and four pulmonary arteries. From the common ventricle at the base of the heart arose a single arterial vessel, forming an aortic arch and continuing as an ascending aorta. At the commencement of the arterial trunk there were three semilunar cusps forming a truncal valve. The right coronary artery arose above the right cusp of truncal valve and the origin of the left coronary artery was situated above the left cusp of truncal valve. The pulmonary arteries arose separately from the left posterolateral aspect of the common arterial trunk (Fig. 4).

According to the presented autopsy findings we diagnosed cor biloculare associated with truncus arteriosus communis. Neither the presence of cor biloculare, nor the persistent truncus arteriosus was diagnosed by the prenatal and postnatal ultrasound examination of the heart. There were no pathomorphological changes present in the other organs. Tracheoesophageal fistula was excluded based on passing a probe through oesophagus and trachea which revealed anatomically correct course and no connection between them. The main bronchi also had a normal course and entered each lung at the hilum. The examination of the lungs showed no signs of congenital respiratory disease. Microscopically we detected hyaline membranes, dilatation of lymph vessels, and a mild amniotic fluid aspiration in what seemed to be the canalicular stage of lung development. Microscopic examination of the myocardial slice taken from the cross section through the common ventricle confirmed absence of interventricular septum. Placenta and extraembryonic membranes were histologically without any pathological findings. The cause of death was cardiopulmonary arrest.

**DISCUSSION**

Cor biloculare with truncus arteriosus communis is an extremely rare congenital cardiac anomaly. Abbott, who was the first to describe and properly study this anomaly reported it occurring 9 times in her series of 1000 cases of congenital cardiac diseases [3].

Cor biloculare represents a congenital cardiac disease in which the heart has only one atrium and one ventricle communicating together by a common atrioventricular valve. This morphological disorder resulting from the undevelopment of interatrial and interventricular septum represents one of the most primitive types of cardiac anomalies [2, 4, 5]. The presence of two-chambered heart alone is very rare, often it is accompanied by other anomalies in the formation of...
the heart as well. In regard to that, Brown (1950) divided cor biloculare to 3 forms: complete form with undivided truncus, complete form with aorta and pulmonary trunk normally divided, and the incomplete form with some septal formation but still a single persistent atroventricular valve [5, 6]. The presented case is an example of the first form.

Truncus arteriosus communis represents a congenital cardiovascular anomaly characterized by a single arterial trunk which arises from the normally developed ventricles, in case of cor biloculare from one common ventricle. This common arterial trunk gives rise to the coronary arteries, as well as the right and left pulmonary artery, and continues to form aortic arch and descending aorta. Classification by Collett and Edwards (1949) distinguishes 4 types of truncus arteriosus communis based on the anatomical origin of pulmonary arteries. In type I, a single short pulmonary trunk originates from truncus arteriosus and gives rise to both pulmonary arteries. Separate, but proximate origins of pulmonary arteries, usually from posterolateral aspect of the common arterial trunk are referred to as a type II, and separate origin of right and left pulmonary artery at some distance from each other as a type III. Type IV is now referred to as a form of pulmonary atresia with ventricular septal defect [7, 8]. Van Praaghs (1965) proposed advanced classification, in which type A1 corresponds with type I of Collett and Edwards, and type A2 includes type II and III. Type A3 refers to the cases where the origin of one pulmonary artery is absent in the common arterial trunk, and the blood supply to that lung is provided by ductus arteriosus or collateral arteries. Type A4 is not defined by the origin of pulmonary arteries, but by the coexistence of the underdevelopment of the aortic arch, including hypoplasia, intrinsic stenosis, or even complete interruption [8, 9]. In our case, truncus arteriosus communis accompanying cor biloculare seemed to be of type II by Collett and Edwards.

Similar cases of cor biloculare with a persistent truncus arteriosus were reported by Tow (1931), Michelson (1943) and Conn et al. (1950). Cordier et al. (1938) also described a heart with single arterial trunk, however they didn’t state whether this vessel was a persistent truncus arteriosus, aorta or pulmonary artery. Michelson (1943) describes truncus arteriosus of type I, with the origin of one pulmonary trunk giving rise to the right and left pulmonary artery, and Tow (1931) is presenting a type III of truncus arteriosus communis, with a separate origins of pulmonary arteries from right and left lateral aspect of the trunk. Conn et al. (1950) in their two cases of cor biloculare with a persistent truncus arteriosus describe the separate origin of pulmonary arteries, however they don’t specify the exact anatomical place of these origins [3, 10, 11].

In our case the common atrium had only one enlarged appendage. Similar finding in case of cor biloculare was reported by Rossman (1942). Also Michelson (1943) describes one (right) atrial appendage with non functioning incompletely developed left appendage [3, 12]. Two appendages are of the more common occurrence, described in cases such as those reported by Popják (1942), Shechter and Meranze (1944), Campbell et al. (1952) and Dhanwate et al. (2015) [2, 5, 13, 14].

Typical characteristic of cor biloculare is a single AV orifice, however Taussing (1947) described an unusual finding of two AV orifices connecting common atrium and common ventricle [5]. Various numbers of cusps in common AV valve has been reported – four [5, 11, 12, 13], three [14], or two [2, 10, 15]. Our case is presenting a common AV valve with three cusps.

Cor biloculare can be associated with anomalous pulmonary venous connection. Some authors described findings of two pulmonary veins opening in the common atrium [12, 13], or even one common pulmonary vein, formed by the union of right and left pulmonary vein before entering common atrium [2, 5].

Case reports of cor biloculare with a separate aorta and pulmonary artery developed describe different anomalies of these great vessels and the systemic blood supply. Common findings were pulmonary artery stenosis or atresia [4, 5, 12, 13] with blood supply to the pulmonary circulation provided by a persistent ductus arteriosus connected to the origin of the right and left pulmonary artery. Some authors reported complete or partial transposition of pulmonary artery and aorta [5, 6, 11, 12, 15]. Two cases included presence of a rudimentary chamber in the common ventricle from which aorta [6] or both aorta and pulmonary artery originated [5]. Nelson and Wells (1948) also reported a presence of the rudimentary chamber, however neither of the great vessels arose from it [15].

Systemic venous anomalies have also been described, such as the absence of inferior vena cava (IVC) [5], common opening of the superior and inferior vena cava placed in the atrial appendage [12], or the presence of bilateral superior vena cava (SVC) with the left SVC entering the common atrium through coronary sinus [2, 5]. Coronary arteries in most cases of cor biloculare originated normally, above the aortic valve, or the semilunar valve of the common arterial trunk. The only unusual finding regarding origin of the coronary arteries was reported by Tow (1931), who in his case described that no coronary vessels arose from the aortic sinuses, but there was a branch of the left pulmonary artery present, which entered the heart muscle [10].

Cor biloculare can also be associated with anomalies of other organs. Situs inversus in various extent is commonly reported. Campbell’s (1952) case has stomach and spleen positioned on the right side, liver was central with peritoneal attachments proving its transposition [5]. Similar findings of partial or
incomplete situs inversus of abdominal organs was reported by Kugel (1932), Rossman (1942), and Conn et al. (1950), almost complete transposition of abdominal organs is mentioned in the case of Nelson and Wells (1948) [5, 11, 12, 15]. Taussing (1947) described a complete situs inversus together with dextrocardia, whereas Shechter and Meranze (1944), Conn et al. (1950) and Yan-bo et al. (2014) have a mention of dextrocardia without any transposition of the abdominal viscera [4, 5, 11, 14]. Another commonly reported finding associated with cor biloculare is asplenia [5, 11, 12]. Combination of cor biloculare with tracheal atresia was reported by McCracken et al. (1964) [6]. Rossman (1942) described malformations of the spine, such as moderate spina bifida of cervical vertebrae, and abnormal development of upper two lumbar and last thoracic vertebrae [12]. Micronathia, cleft palate, glossoptosis, hypertelorism, and left lower limb deformity associated with cor biloculare reported by Dhanwate et al. (2015) could be suggestive of some complex syndrome [2]. The presented case was without any traces of anomalies of other organs. Cor biloculare with truncus arteriosus communis belongs to the group of cyanotic congenital heart diseases. Blood from both systemic and pulmonary veins is entering the common atrium, therefore the mixing of oxygenated and deoxygenated blood occurs. Through single atrioventricular valve the blood is passing into the common ventricle. Due to the presence of single arterial trunk through which the blood is entering systemic and pulmonary circulation, further mixing occurs [2, 4, 16]. Most babies born with cor biloculare usually die shortly after the birth. Longest survival reported so far has been 44 years [4]. Survival in the reported cases of cor biloculare associated with truncus arteriosus communis has not exceeded 5 months [10]. In the presented case the child born prematurely, at the 23th week of pregnancy dies approximately 34 hours after the birth due to cardiopulmonary arrest.

The presence of arterial septal defect was already confirmed to have a genetic origin, and also the presence of other accompanying organ abnormalities mentioned in case reports of cor biloculare in the literature would suggest a certain genetic condition [17]. However no genetic tests were performed after the autopsy to rule out or confirm this suggestion.

Presented case of cor biloculare reports its rarely reported association with truncus arteriosus communis, without other organ malformations, as well as any abnormalities visible during external examination.

**Conflict of interest.** Authors declare that there is no conflict of interest concerning this article.

---

**References**