



## Case report

## Case report of rare congenital cardiovascular anomalies associated with truncus arteriosus type 2



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## ARTICLE INFO

## Keywords:

Truncus arteriosus  
Fetal heart  
Cardiovascular diseases  
Heart defects  
Congenital

## ABSTRACT

Truncus arteriosus (TA) is a very rare congenital anomaly with complex cardiovascular anatomy and high lethality also due to severe associated anatomical variants and pathologies. As TA has a massive impact on the survival of a newborn and usually has to be surgically treated. Thus, it is of high importance to understand this congenital cardiovascular disease and associated complications, to improve life expectancy and outcome of these patients. We recently came across a newborn female patient with a rare complex case of persistent TA type 2 associated with further complex cardiovascular anomalies, who received a contrast enhanced CT scan on the 3rd day post-partum, showing complex cardiovascular abnormalities that were ultimately incompatible with life.

## 1. Epicrisis

## 1.1. Day of birth and the first clinical examination

We report the case of a female newborn with birth by sectio after frustrated induction due to macrosomia associated with a poorly managed dietary gestational diabetes mellitus. Good postnatal adaptation of the newborn was present with an APGAR score of 9/10/10. The baby was in a good general condition, crying vigorously, showed good spontaneous motor function and the fontanel was soft and in level. Skin color was rosy without icterus or exanthema. The newborns cardiac action was rhythmic with a good recapillarization time (<2 s). All laboratory values were physiological. Both lungs were well ventilated with eupnea. Abdomen was soft without palpable resistance and regular bowel sounds. Good and laterally equal spontaneous motor activity was reported. Due to already prenatally known suspicion of hypoplastic left heart syndrome, therapy with Minprog (Alprostadil, prostaglandin 1 inhibitor) (5 ng/kg/min) was started immediately. Preductal saturation was fluctuating between 70 and 85%, without O<sub>2</sub> administration. Vitamin K was later applied i.v. on the normal ward.

## 1.2. Echocardiography on the 2nd day post partum

In echocardiography mitral atresia with hypoplastic left ventricle was reported. Further, the right ventricle was hypertrophied with unremarkable function. The tricuspid valve showed no stenosis or regurgitation on color Doppler sonography. From the right ventricle a large vessel arised from which the pulmonary arteries originated, which was being noted as a truncus arteriosus (TA). Furthermore, the large brachial and cephalic arteries originated relatively distally from the outer curvature of this "truncal vessel". The TA was connected to the descending aorta via a wide-open ductus arteriosus Botalli (PDA). An aortic valve or ascending aorta could not be visualized. Highly suspected coronary anomaly with origin of the coronary arteries from the truncus brachiocephalicus was reported. Atrial septum defect (ASD) II with unaccelerated left-to-right shunt was detected. There was no evidence of malapposition of pulmonary veins.

## 1.3. Computed tomography angiography (CTA) of the heart on 3rd day post-partum

CTA showed a persistent TA type II (Figure 1A and B), a hypoplastic left ventricle, ventricle-septum-defect (VSD), ASD, and mitral atresia,

This article is a part of the "Advanced Imaging in Cardiovascular Diseases" Special issue.

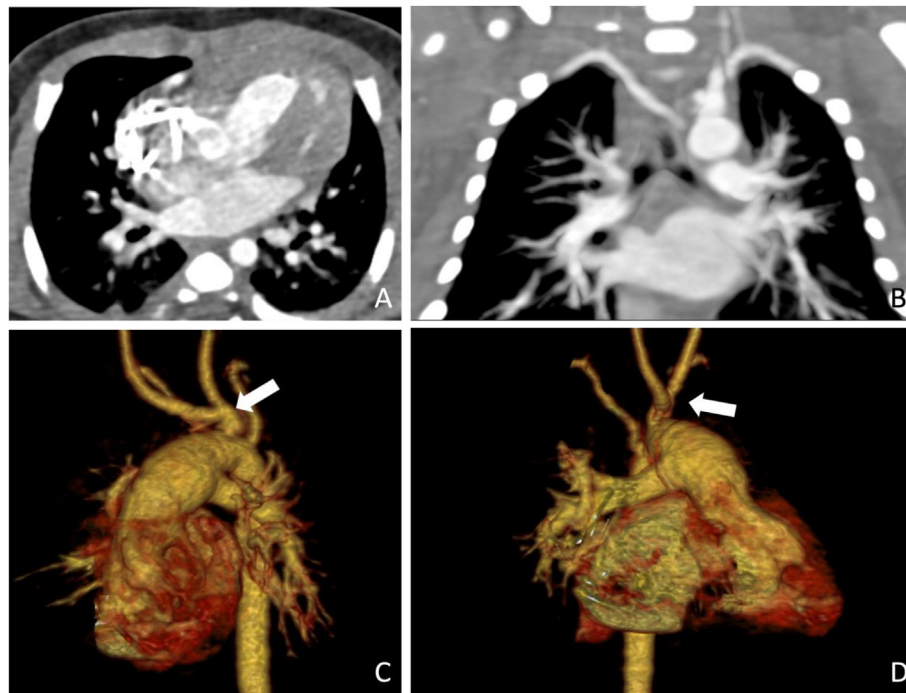
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<https://doi.org/10.1016/j.heliyon.2022.e11033>

Received 15 February 2022; Received in revised form 20 June 2022; Accepted 6 October 2022

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**Figure 1.** Cardiac CT with intravenously injected contrast agent demonstrated the massive hypoplasia of the left ventricle with collapse of the lumen and pellotation by the ventricular septum in axial view (A). In coronal plane, the aberrant right subclavian artery originating as a lusorian artery is visible, which originates from the truncus separately from the truncus bicaroticus (B). In the volume-rendering-technique (VRT) reconstructions the truncus bicaroticus is depicted (C and D, arrows).

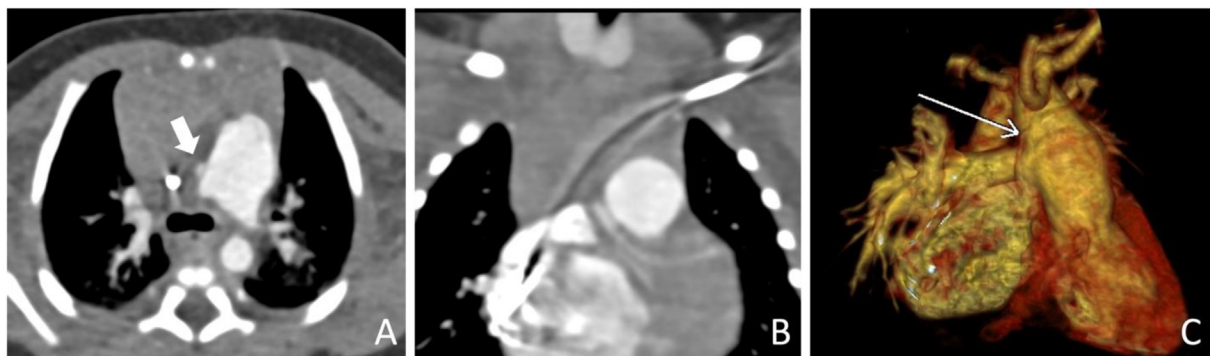
among a lusorian artery (Figure 1 C and D). Furthermore, a single main coronary artery (Figure 2A) originated from a truncus bicaroticus, just caudal to the origin of the right common carotid artery with course caudally along the right-sided TA and branching into three coronary arterial branches (Figure 2B and C). The lusorian artery showed a typical course. Consecutively, hypoplastic left heart syndrome with mitral atresia was diagnosed with agenesis of the ascending aorta. Systemic perfusion was sustained by PDA.

#### 1.4. Echocardiography as of 27th day post-partum

The atrial septum was reported as slightly wider, with a left-right shunt and turbulence on color Doppler sonography. The mean gradient was 15 mmHg. The left atrium appeared less dilated, compared to initial echocardiography. Good function of right ventricle was reported. The truncal valve showed minimal central insufficiency with regurgitation. The descending aortic arch showed no stenosis and the PDA a good flow of 2 ml/s. Only slight diastolic return flow was described. Flow in the TA was good in the systole and showed minimal reflux in the diastole.

## 2. Clinical course

On admission, echocardiographic evidence of hypoplastic left heart syndrome with agenesis of the ascending aorta with PDA-dependent systemic perfusion was reported. Therapy with alprostadil was initiated and a central venous catheter implemented. In addition, start of antibiotic therapy with ampicillin and cefotaxime was performed due to persistent fever and rising infection parameters and detection of a resistant *Streptococcus cristatus* in the throat swab. Antibiotic therapy was switched to vancomycin and meropenem because of persistent symptoms on the third day post-partum. Antibiotic therapy was continued until 15<sup>th</sup> day post-partum. On the 21st day post-partum fever occurred again, the central venous catheter was removed and antibiotic therapy was performed for 7 more days. Overall, stable cardiorespiratory conditions, unremarkable monitor monitoring, adequate drinking behavior, and good weight gain were noted. From our point of view and after interdisciplinary demonstration and discussion with the colleagues at the pediatric cardiac center in BLINDED, there were no reasonable therapy options due to the complex cardiovascular anatomy. After



**Figure 2.** Cardiac CT with intravenously injected contrast agent demonstrated the origin of a singular coronary artery from the truncus arteriosus (TA) in axial and coronal plane (A, arrow; B) as well as in the volume-rendering-technique (VRT) (C, arrow). Further, the origin of two separate pulmonary arteries directly originating from the TA is visible. The beam hardening artefacts in the left subclavian vein and upper caval vein are due to a central catheterization and injected contrast agent.

presentation of the patient and consultation with the parents at the Pediatric Heart Center in BLINDED, the family could not be offered heart transplantation listing. To allow palliative care at home without Minprog therapy, a cardiac catheterization with implantation of an 8 × 18 mm stainless-steel drug eluting stent into the TA and a Rashkind maneuver for restrictive atrial septum defect was performed on the 44<sup>th</sup> day post-partum. After this, therapy with alprostadil could be stopped and the central vein catheter was removed. Discharge to outpatient care followed. After two months the child unfortunately ceased, due to the severity of the congenital heart anomalies. As cause of death heart failure due to cardiac insufficiency and consecutive cerebral hypoxia as well as pulmonary edema were stated. Informed consent of the parents was obtained to publish this case report.

### 3. Discussion

TA is a rare congenital cardiac anomaly, where a single artery originates from the ventricular outflow tract to provide pulmonary, coronary, and systemic circulation [1, 2, 3]. In contrast to the aortic valve in a physiological heart, the TA only has a single semilunar valve, often causing regurgitation or stenosis [1, 4]. TA is often accompanied by a non-restrictive anterior VSD which grants the passage of blood from the left to the right ventricle, as the left ventricle is commonly hypoplastic and with little to no function [5]. The TA is classified in different types (Type 1–4 in Collett and Edwards or Van Praagh classification) depending on how many structures originate directly from it. Like in our case, patients often have additional anomalies of the vascular anatomy, such as a single coronary artery (8–14% of cases) [6, 7, 8]. The TA is also associated with stenosis of the pulmonary arteries in 2–10%, which may originate as two separate arteries from the truncus - like in our patient - or together as a common truncus pulmonalis [1]. In this context CT is a more accurate modality compared to ultrasound alone for visualization of the complexity of anomalies and to plan potential surgery [9, 10, 11, 12]. Thus, additionally to sonography, on the 3rd day post-partum we performed a CT scan with intravenously injected contrast material to further analyze the anatomical situation and to plan possible therapy. Exemplary, in our case the origin of the main coronary artery was only correctly diagnosed in CTA, not in the initial echocardiography. In our patient we also found a lusorian artery as well as a truncus bicaroticus and an ASD [5]. Atresia of the mitral valve and the hypoplastic left heart of our patient correspond with the low to missing functionality of the left heart, as the body circulation is solely sustained by the hypertrophied right ventricle. The right heart is hypertrophied as it compensates the insufficient left heart function and provides for the pulmonary, as well as to the systemic circulation. Surprisingly, the initial condition of the newborn was good with an APGAR score of 9/9/10. This might be explained by the fetal circulation, which was not compromised by the heart anomalies until birth [8]. In a healthy newborn, the first screams and breaths inflate the lungs and build up enough resistance from the pulmonary arterial system to close the oval foramen, thus dividing the left and right hearts circulation. Further, the peri- and post-partial expression of prostaglandins leads to obliteration of the ductus arteriosus Botalli within the first days after birth. In our patient, the atrial and ventricular septal defects were lifesaving after birth, as otherwise the blood circulation of the heart would have been compromised, leading to peripartur death, as the pulmonary veins still feed into the left atrium. From the left atrium the oxygenated blood would usually get pumped to the body by the left ventricle, which in our patient is hypoplastic. Thus, the oxygenated blood needs to be transported from the left to the right atrium again. The oxygenated and unoxygenated blood gets mixed in the right atrium and ventricle and then pumped through the TA to the systemic and pulmonary circulation. This mixed arteriovenous semi-oxygenated blood was sufficient at the time of birth and led to oxygenation problems only later.

Our patient therefore received a percutaneous cardiac intervention with implantation of an 8 × 18 mm stainless-steel drug eluting stent into

the TA and a Rashkind maneuver for restrictive ASD on the 44th day post-partum, aiming to sustain circulation [13, 14], as in interdisciplinary consensus a curative approach was not possible. A curative option for minor cases of cardiac anomalies associated to a TA is a Norwood or Fontane I or II surgery, in which the pulmonary artery is reconnected to the right heart using a graft and VSD is closed [13, 14]. As this surgery is very difficult and the outcome often is not optimal, many peri- and postoperative complications occur [13, 14]. Unfortunately, our patient was not stable enough to receive full anaesthesia and surgery, also the anatomical variant with only one common coronary artery made a curative approach impossible.

Overall, our patient was in a palliative situation straight after birth, because no reasonable therapeutic options were available due to the complexity of the heart anomalies. If the child had remained clinically stable, another cardiac catheterization with intraluminal pulmonary artery banding might have been considered. Yet, in our patient this was not possible, as the congenital hypoplasia of the left ventricular outflow tract could not be compensated. In conclusion, our patient died after only two months due to the heart's inability to sustain systemic, pulmonary and coronary circulation, as well as due to massive heart insufficiency due to the massive valve regurgitation of the TA. The consecutive hypertension in the dilated left atrium was causing pulmonary edema. In the end, all these pathologies combined were not compatible with life.

### Declarations

#### Author contribution statement

All authors listed have significantly contributed to the investigation, development and writing of this article.

#### Funding statement

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

#### Data availability statement

Data included in article/supp. material/referenced in article.

#### Declaration of interest's statement

The authors declare no conflict of interest.

#### Additional information

No additional information is available for this paper.

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