

CASE REPORT

Persistent Common Arterial Trunk in an Adult Presenting in the Emergency Room as Severely Decompensated Heart Failure

Zsuzsanna Szöke, András Suciu, Géza Jeszenszky, Piroska György

Clinic of Cardiology, Municipal Hospital, Gheorgheni, Romania

ABSTRACT

Truncus arteriosus (TA) or common arterial trunk is a rare malformation, accounting for 0.21 to 0.34% of congenital heart diseases, which, if left untreated, leads to increased mortality rates. The condition is characterized by the presence of a unique arterial trunk that overrides the interventricular septum. Despite an overall poor outcome, few subjects present in emergency settings with signs suggestive for pulmonary arterial hypoplasia and associated heart failure. We report the case of a 31-year-old female patient who had been previously diagnosed with pulmonary atresia and severe scoliosis as an infant, presenting in the emergency department with clinical signs of decompensated heart failure which were demonstrated to be attributable to the severe cyanogenic heart malformation and were reversible after initiation of appropriate therapeutic measures.

Keywords: common arterial trunk, MAPCA, interventricular septum defect

ARTICLE HISTORY

Received: July 11, 2017

Accepted: August 19, 2017

CORRESPONDENCE

Andras Suciu

Bul. Lacu Roşu nr. 16

535500 Gheorgheni, Romania

Tel: +40 266 364 008

E-mail: suciuandrasjr@yahoo.com

INTRODUCTION

Congenital cardiac defects (CCD) have an incidence ranging between 4 and 10 cases in every 1,000 alive births in the United States and 6.9 per 1,000 in Europe.¹ CCD consist of an abnormal development of various cardiac and vascular structures during the intrauterine life, which leads to subsequent anatomic and hemodynamic consequences requiring complex surgical and interventional therapies. These critical defects have been associated with intrauterine mortality and high rates of deaths during childbirth, infancy, or childhood, as well as with an increased morbidity and mortality in pediatric and adult populations. Furthermore, in addition to a decreased quality of life,

CCD imply increased hospitalization rates and elevated healthcare costs.²

The development of complex imaging methods for the diagnosis of these critical conditions and complex surgical and interventional therapies addressed to such patients, has led to a substantial improvement in their life expectancy and in the associated survival rates. In recent years, the significant progress encountered in surgical, anesthetic, and perioperative procedures resulted in a significantly improved survival of patients with CCD, the age of death shifting from infancy and pediatric age towards adulthood. Despite of this improvement in the complex management of CCD patients, the risk for morbidity and

mortality in the CCD population remains high, mainly due to the associated heart failure and malignant arrhythmias.³

Truncus arteriosus (TA) or common arterial trunk is a rare malformation, accounting for 0.21 to 0.34% of congenital heart diseases, which, if left untreated, leads to increased mortality rates. The condition is characterized by the presence of a unique arterial trunk that overrides the interventricular septum, supplying both the pulmonary and systemic circulations, as well as the coronary arteries.⁴ Depending on the origin of the pulmonary arteries, there are four types of TA according to the Collett-Edwards anatomical classification system: type I, in which a short pulmonary trunk arises from the TA, dividing into the two pulmonary arteries; type II, in which both pulmonary arteries separate from the posterior wall of the TA; type III, where the pulmonary arteries arise from the lateral aspects of the TA; and type IV, in which the TA is overriding a large ventricular septum defect and is considered a more severe type of the tetralogy of Fallot.^{5,6}

If left untreated, common arterial trunk presents a severely impaired prognosis and high mortality rates, causing early congestive heart failure and pulmonary hypertension with cyanosis in most of the cases. Associated cardiac malformations include right-sided or interrupted aortic arch, aortic coarctation, atrioventricular septum defects, and common atrioventricular junction, as well as coronary artery anomalies that may be related to an anomalous origin or trajectory.⁷

Despite an overall poor outcome in case of uncorrected TA, due to severe pulmonary arterial hypertension caused by pulmonary overflow and refractory heart failure, few subjects present in emergency settings with signs suggestive for pulmonary arterial hypoplasia and associated heart failure.⁸

We report the case of a 31-year-old female patient who had been previously diagnosed with pulmonary atresia and severe scoliosis as an infant, presenting in the emergency department with clinical signs of decompensated heart failure which were demonstrated to be attributable to the severe cyanogenic heart malformation and were reversible after initiation of appropriate therapeutic measures.

The patient agreed to the publication of her data and the institution where the patient had been admitted, approved the publication of the case.

CASE REPORT

A 31-year-old woman presented in the emergency department with severe dyspnea and orthopnea, cyanosis, and

an overall impaired functional capacity. The medical history revealed that she had been diagnosed during infancy with pulmonary atresia, left ventricular septum defect, bronchopulmonary and arterial collateralization, by undergoing echocardiographic and angiographic examinations. The patient had not undergone surgical treatment due to her severe scoliosis and was discharged with pharmacological therapeutic recommendations.

At the age of 14 she was admitted in England for further investigations due to moderately restricted exercise tolerance and had undergone cardiac catheterization for evaluation of the cardiac pressures and angiographic examination that had revealed three major aorto-pulmonary collateral arteries (MAPCAs) that supply the right lung at the level of the descendant aorta, while the left lung had no visible flow. Upon injection in the left pulmonary vein, the angiography revealed a retrograde filling of a central pulmonary artery within the hilum of the left lung, with a diameter of approximately 3 mm. Again, she had not undergone surgical correction of the cardiac malformation due to the high anesthetic risk and severe scoliosis. Over the following years, the patient had been admitted several times and had been receiving oxygen therapy at home in the last three years before the present admission.

At presentation in the emergency room, the patient exhibited severe sinistro-convex dorsal and compensatory lumbar dextro-convex scoliosis and was bound to a wheel chair, weighing 40 kilograms. She presented central and peripheral cyanosis, and clubbing of the fingers. There were no abnormal pulmonary sounds, and upon cardiac auscultation she presented a continuous murmur. The blood pressure was 100/65 mmHg, heart rate 100 beats per minute and the respiratory rate 20 breaths per minute with an oxygen saturation of 60% in the ambient air. The electrocardiographic examination revealed a normal sinus rhythm, right axis deviation, and right ventricular hypertrophy.

The current symptoms suggested a worsening of the heart failure, and the patient underwent an echocardiographic examination that revealed a concentric right ventricular hypertrophy, the presence of perimembranous interventricular septum defect with a right-to-left shunt, and a left aortic arch (Figure 1, panels A, B, C, and D).

An ECG-gated 128-multislice cardiac computed tomography performed in emergency showed that the common arterial trunk, which was measured as having a diameter of 4.79 cm (Figure 2A), was overriding the perimembranous interventricular septum defect (14/24 mm – Figure 2B). The descending aorta presented 3 MAPCAs with tortuous course, two of them originating at the level of the right, and one from the left aortic wall, all supplying the

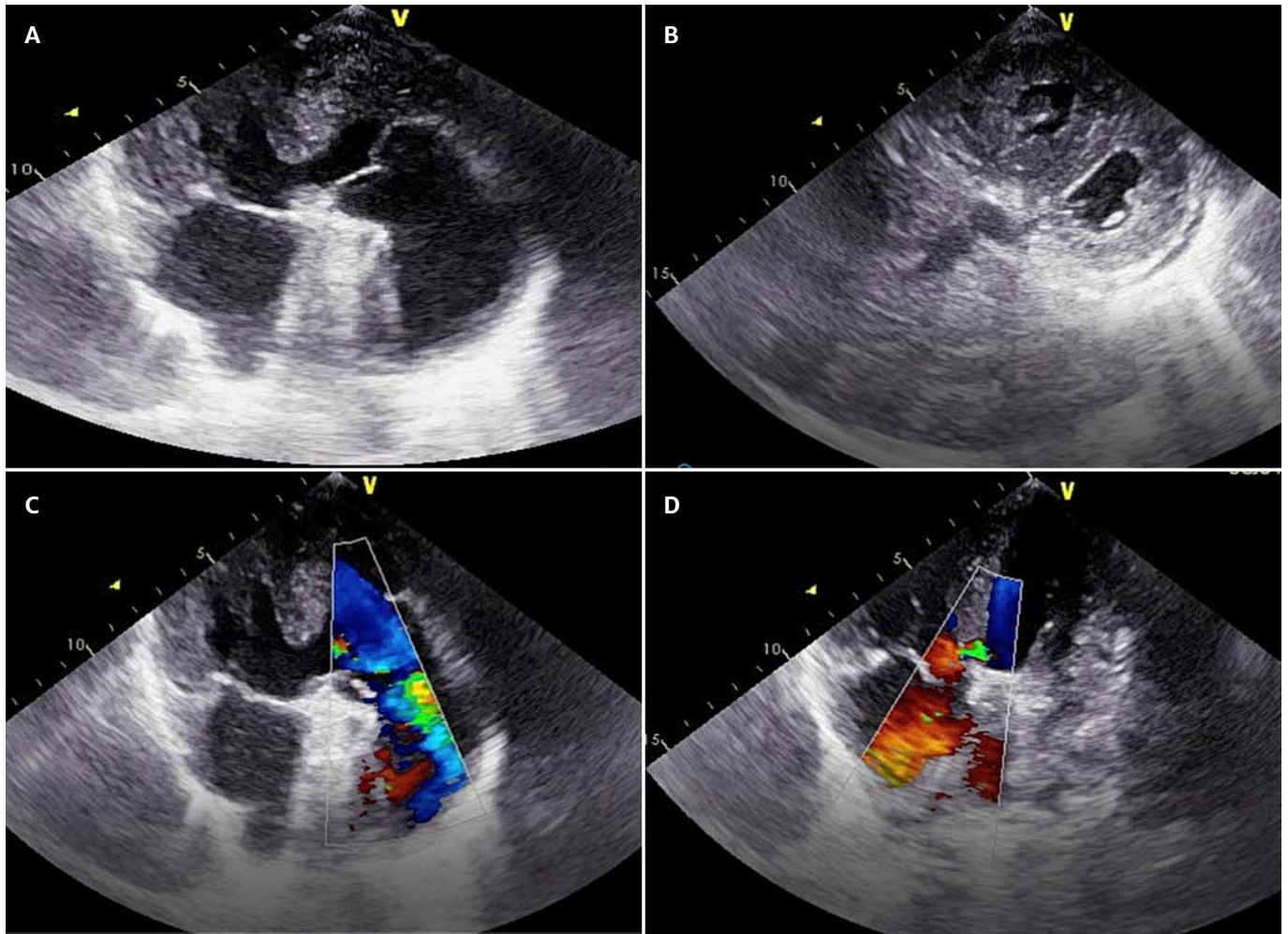


FIGURE 1. Transthoracic echocardiography. **A** – Apical four-chamber view with the ventricular septum defect and the left positioning of the aortic arch. **B** – Apical four-chamber view with Color Doppler showing the turbulent flow through the common arterial trunk. **C** – Parasternal short axis view revealing the right ventricular hypertrophy. **D** – Apical four-chamber view with Color Doppler showing the right to left shunt through the interventricular septum defect.

right lung (Figure 3A, 3B). As for the blood supply to the left lung, there were a few filiform aorto-pulmonary collateral arteries that emerged from the anterior wall of the descending aorta (Figure 3C). According to the CT examination, the patient was diagnosed with common arterial trunk, being categorized as a Collett-Edwards class IV (Figure 4A, 4B).

The therapeutic management included pharmacological treatment with digoxin, diuretics, and ambulatory oxygen therapy, and the patient was closely monitored, being discharged with significant symptoms relief.

DISCUSSIONS

If left uncorrected, TA presents a poor prognosis, with a mortality rate of 80% in the first year of life, and unrepaired cases that survive up to adulthood are rare. TA is

an extremely rare CCD, being characterized by a common vessel that emerges from both ventricular cavities on top of a large septum defect and feeds all three circulations (coronary, systemic, and pulmonary).⁹⁻¹²

MAPCAs are vessels developed in the intrauterine life that regress after proper growth of the pulmonary arteries. In cases where the pulmonary blood flow is impaired, such as tetralogy of Fallot or pulmonary atresia, MAPCAs do not regress, but continue to develop as the main arteries supplying the pulmonary circulation.¹³ Depending on the origin of the MAPCAs, the TA is classified in four types.⁵ The presented case was included in type IV, as she presented with the origin of the aorto-pulmonary branches at the level of the descending aorta. Type IV TA, the rarest of the four categories, is also referred to as pseudo-truncus arteriosus, as it is difficult to differentiate from the tetralogy of Fallot with pulmonary valve atresia.^{5,14}

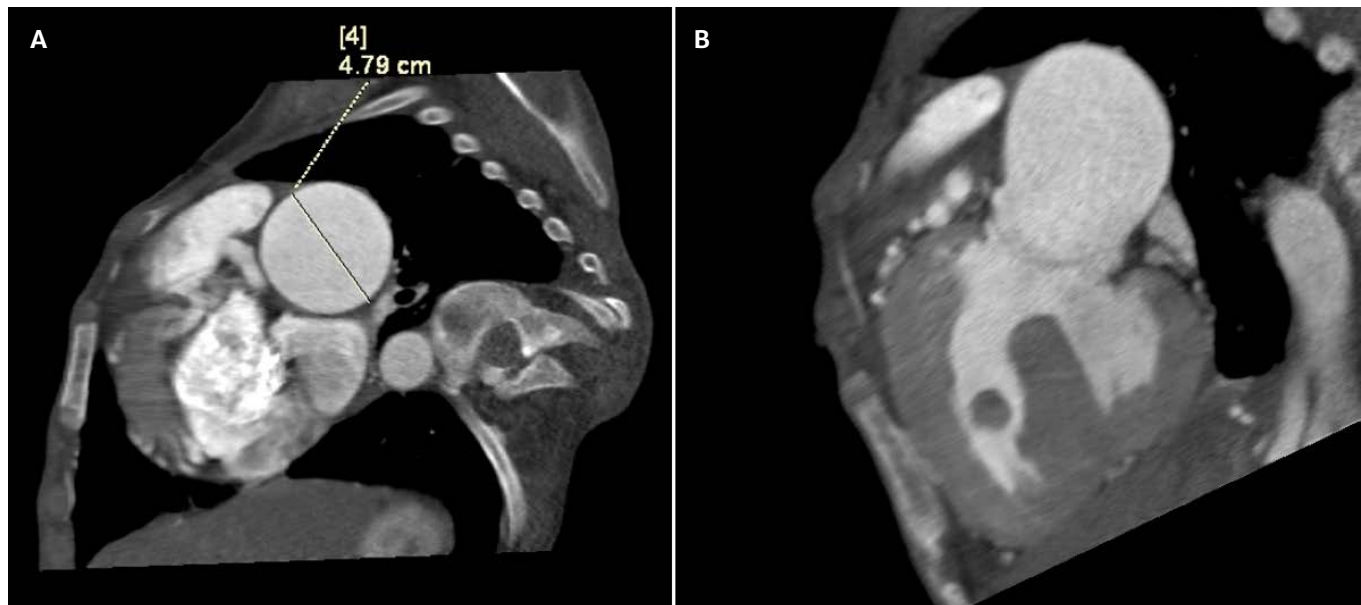


FIGURE 2. Cardiac 128-slice CT. **A** – Common arterial trunk. **B** – Interventricular septum defect.

The large ventricular septum defect is a malformation associated with the TA, and the direction of the shunt depends on the vascular resistance ratio between the systemic and pulmonary circulation. The left-to-right shunt is usually present in early stages of life, and it leads to congestive heart failure in TA infants. In time, due to increased pressures in the arterial pulmonary circulation, vascular remodeling with intimal fibro-elastosis leads to development of severe pulmonary hypertension with reversal of the shunt direction (Eisenmenger syndrome). In cases of TA it is the main reason for survival through adult life if left surgically uncorrected.¹⁵

Imaging modalities in CCD include echocardiography, which in TA allows the assessment of: (1) the common arterial trunk; (2) the TA valve, which in this case was tricuspid; (3) the associated valvular stenosis or regurgitation; and (4) the shunt direction and pressure gradients.¹⁶ Multislice computed tomography (MSCT) is a noninvasive imaging method that can offer both visualization of cardiac structures and assessment of coronary, pulmonary, and systemic vasculature. At the same time, this technique allows visualization of extracardiac structures, including the lungs, airways, and spine.^{16,17} In the presented case, the 128-slice CT allowed proper visualization and measurement of the ventricular septum defect, the anatomical

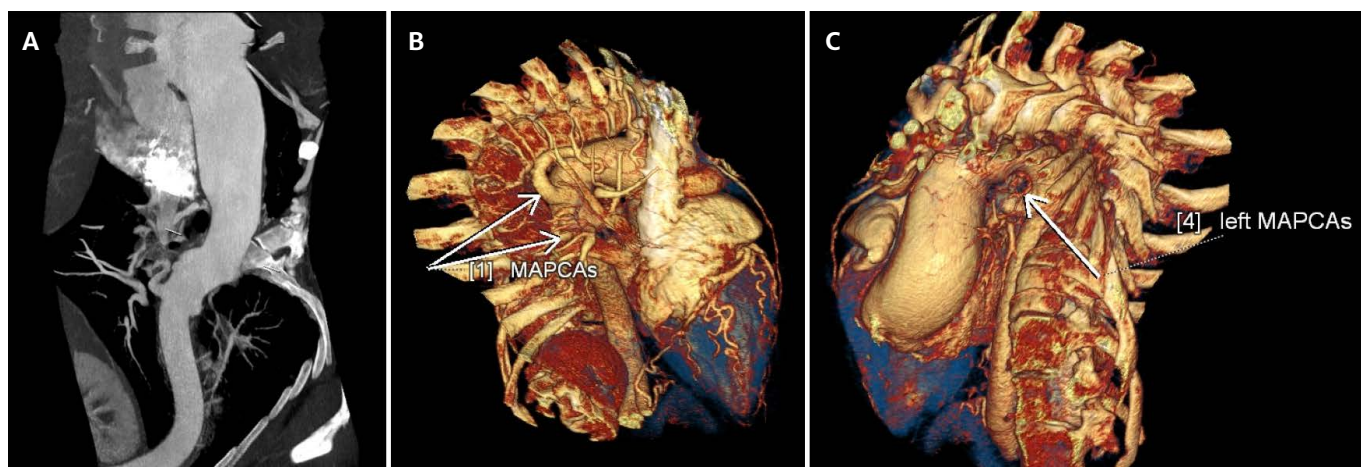


FIGURE 3. CT angiography of the descending aorta. **A** – The three major aorto-pulmonary collateral arteries for the right lung. **B** – Collateral aorto-pulmonary branches for the right lung – 3D volume rendering. **C** – Filliform MAPCAs to the left lung – 3D volume rendering.

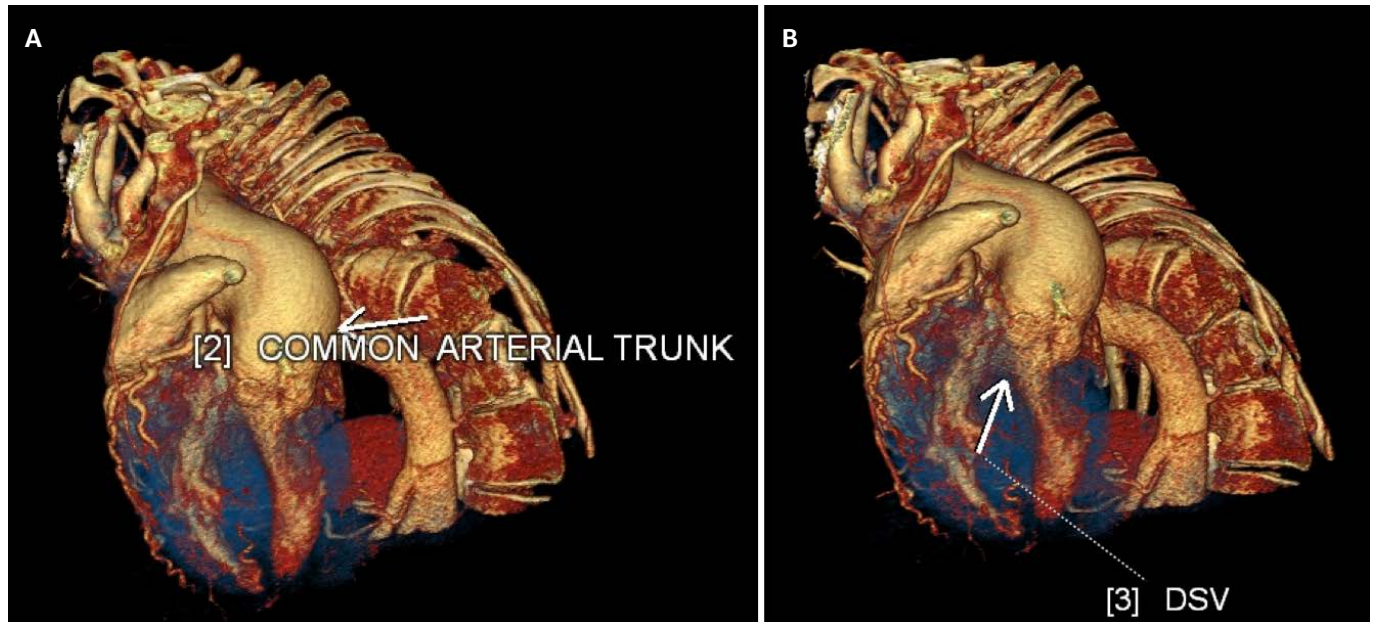


FIGURE 4. Cardiac CT with 3D reconstruction of the heart and malformations. **A** – Common arterial trunk. **B** – Ventricular septum defect

illustration of the MAPCAs, as well as an accurate visualization of the scoliotic dorsal spine that caused deformity of the thoracic cavity and restrictive ventilatory dysfunction that contributed to the increased pulmonary pressures and decreased tissue oxygenation.

Despite the overall general condition of the patient, the lack of surgical correction and severely decreased oxygen saturation (less than 60% in the ambient air), she managed to lead a quasinormal life and has graduated from two universities. The main particularity of this case report is that it shows the natural evolution of a complex cyanogenic congenital heart defect that has not been operated.

CONCLUSIONS

The case report underlines the importance of noninvasive cardiac imaging for achieving an accurate depiction of adult survivors with uncorrected cyanogenic congenital cardiac defects. Common arterial trunk is a rare congenital heart defect with high mortality rates if left surgically untreated. However, this case illustrates that a proper management of this severe condition in emergency settings and also during the follow-up period, makes possible to overcome the odds of a short lifespan and improve the prognosis of these critical patients.

CONFLICT OF INTEREST

Nothing to declare.

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