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Case Report

A rare case of truncus arteriosus Van Praagh type A3: Prenatal diagnosis and postnatal management

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ABSTRACT

Truncus arteriosus (TrA) type A3, according to the Van Praagh (VP) classification, cannot be classified under the Collett and Edwards (C–E) system. In this rare anomaly, postnatal closure of the ductus arteriosus can cause unilateral pulmonary artery obstruction; hence, prenatal diagnosis and early confirmatory postnatal diagnosis are important. This case was referred to our hospital for suspected fetal heart disease at 29 weeks' gestation. TrA C–E type 1 was diagnosed by fetal echocardiography, with a right-sided aortic arch, absent inferior vena cava, and azygos continuation. The neonate was delivered vaginally at 41 weeks' gestation. Postnatal echocardiography showed a right-sided aortic arch with the right pulmonary artery originating from the common arterial trunk and the left pulmonary artery originating from the brachiocephalic artery. The diagnosis was TrA VP type A3, with a right-sided aortic arch and left-sided ductus arteriosus. Patency of the left-sided ductus arteriosus was maintained with prostaglandin E1.alpha-cyclodextrin. Right pulmonary artery banding was performed 3 days after birth. The Rastelli procedure was performed when the patient was 2 months old and weighed 4.2 kg. Delayed diagnosis of VP type A3 can cause unilateral pulmonary artery disconnection; hence, timely and accurate diagnosis is warranted to ensure stable disease management.

<**Learning objective:** Delayed diagnosis of truncus arteriosus (TrA) Van Praagh (VP) type A3 can cause unilateral pulmonary artery disconnection; thus, we should consider both Collett and Edwards and VP classifications, including a ductus arteriosus contralateral to the aortic arch, during TrA diagnosis. Furthermore, to prevent the risks associated with an open-heart surgery in the neonatal period in such cases, pulmonary blood flow can be controlled by maintaining the ductus arteriosus with prostaglandin E1.alpha-cyclodextrin and unilateral pulmonary artery banding.>

Introduction

Truncus arteriosus (TrA) is a rare disease that can be classified using the Collett and Edwards (C–E) or Van Praagh (VP) classification systems. VP type A3 involves the absence of a unilateral proximal pulmonary artery and the presence of ductus arteriosus or collateral arteries, which supply the lungs in the absence of a pulmonary artery branch from the truncus [1]. This type cannot be classified under the C–E classification system, and there are almost no case reports of its prenatal diagnosis. Moreover, postnatal closure of a ductus arteriosus can cause unilateral pulmonary artery disconnection; hence, prenatal diagnosis and early confirmatory postnatal diagnosis are important.

We report our experience of a rare case of VP type A3 with a right-sided aortic arch and left-sided ductus arteriosus. After the neonate was delivered, the patency of the left ductus arteriosus was maintained by infusion of prostaglandin E1.alpha-cyclodextrin (PGE_1-CD), and right pulmonary artery banding was performed, until the patient was considered fit to undergo the Rastelli procedure. We report the fetal echocardiographic findings and postnatal course of the case.

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Case report

A 35-year-old pregnant woman was referred to our hospital at 29 weeks' gestation, for suspected fetal heart disease.

Fetal echocardiography showed one great vessel overriding the ventricular septum (Fig. 1A, B). The main pulmonary artery and right pulmonary artery originated from the ascending part of this vessel, thereafter traveling in the left posterior direction. The origin of the left pulmonary artery was not clearly seen; however, the blood flow toward the left lung was seen originating from the vicinity of the main pulmonary artery (Fig. 1C, D). Based on the above findings, the diagnosis was TrA C–E type 1 with a right-sided aortic arch. There was no stenosis or regurgitation at the common arterial trunk valve. The inferior vena cava was absent with azygos continuation, and left isomerism was suspected.

A female neonate was delivered vaginally at 41 weeks' gestation, with a body weight of 2890 g, Apgar score of 8/9 points, heartbeat of 170 beats per minute, respiratory rate of 40 breaths per minute, blood pressure of 62/31 mmHg, and SpO_2 of 96%. Echocardiography showed a right-sided aortic arch with the right

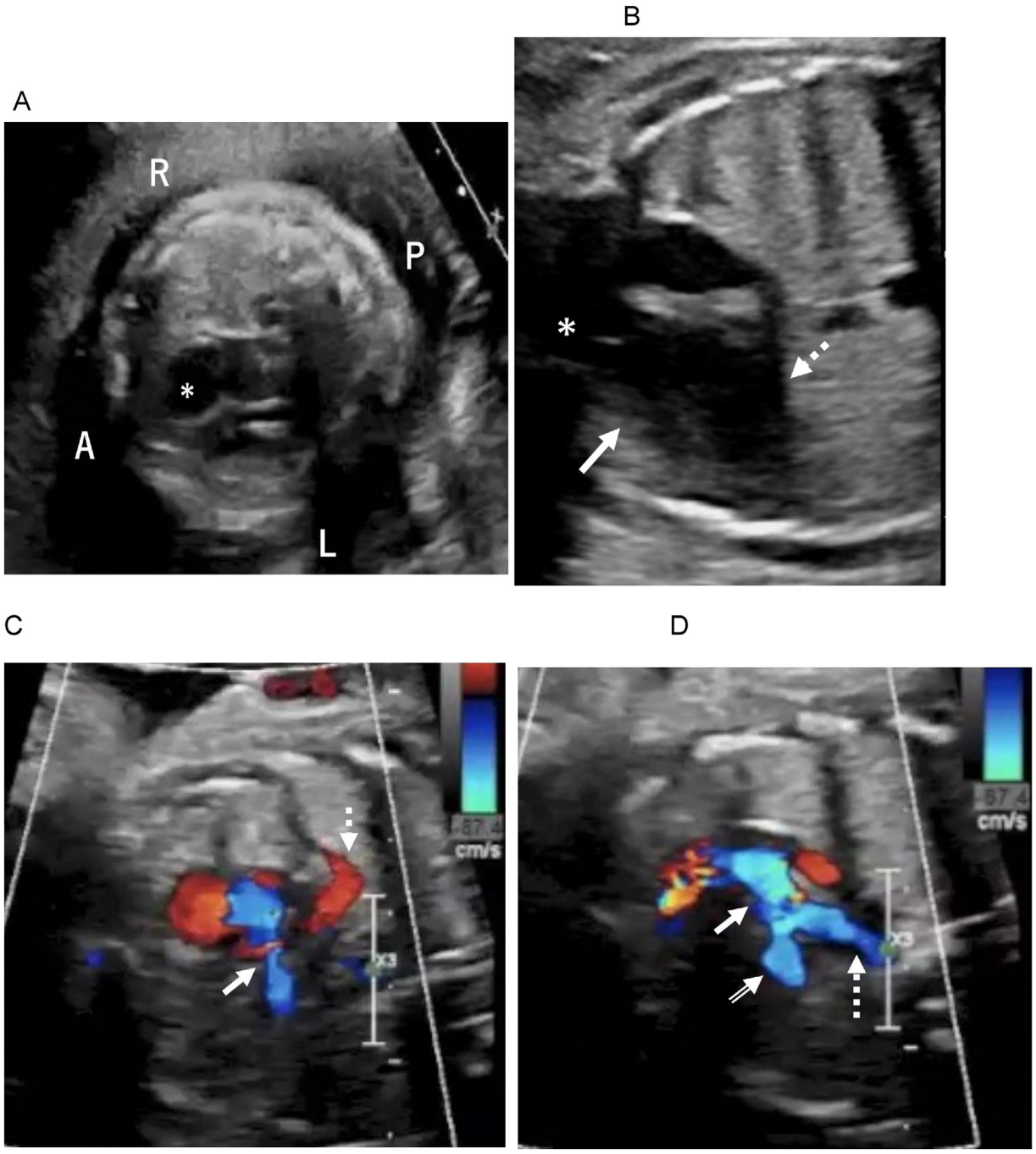

Fig. 1. Fetal echocardiogram at 29 weeks of gestation. (A, B) Image of the common arterial trunk. *Indicates the common arterial trunk, single arrow indicates the right ventricle, and dotted arrow indicates the left ventricle. (C) Origin of the right pulmonary artery. The dotted arrow indicates the right pulmonary artery, and the single arrow indicates the brachiocephalic artery. (D) Origin of the left pulmonary artery. The single arrow indicates the brachiocephalic artery, double-line arrow indicates the left subclavian artery, and dotted arrow indicates the left pulmonary artery.

pulmonary artery originating from the common arterial trunk and the left pulmonary artery originating from the brachiocephalic artery. The postnatal diagnosis was TrA VP type A3 with a right-sided aortic arch and a left-sided ductus arteriosus. Contrast-enhanced computed tomography confirmed the diagnosis (Fig. 2A–C), and left isomerism was diagnosed based on a multi-lobulated spleen (Fig. 2D), bilateral left atrial appendage, and two lobed lungs bilaterally.

At 6 h after birth, the SpO_2 had declined to 86%, and stenosis of the ductus arteriosus was observed at the left pulmonary artery origin; hence, PGE_1-CD 10 ng/kg/min was initiated. In addition, right pulmonary artery banding was performed 3 days postnatally, to restrict high pulmonary blood flow. The right pulmonary artery before banding measured 4.0 mm in diameter, and the circumferential size of the banding was 10 mm. The diastolic blood pressure of the patient increased by 5 mmHg and the SpO_2 decreased by 5% during banding.

Echocardiography showed a peak flow velocity of 3.0–3.5 m/s at the banding site, which meant that the peripheral right pulmonary arteries were prevented from progressing to pulmonary hypertension. With a target SpO_2 of 75–85%, the PGE_1-CD dose was adjusted from 4 to 10 ng/kg/min, to obtain a suitable stenosis at the ductus arteriosus. Echocardiography showed a peak flow velocity of 2.5–3.5 m/s at the left ductus arteriosus. As the patient gained weight, the peak flow velocity at the site of right pulmonary artery banding increased to 4.4 m/s, thereby indicating that stenosis at the banding site had relatively progressed. SpO_2 had decreased to approximately 70%; hence, the Rastelli procedure was performed

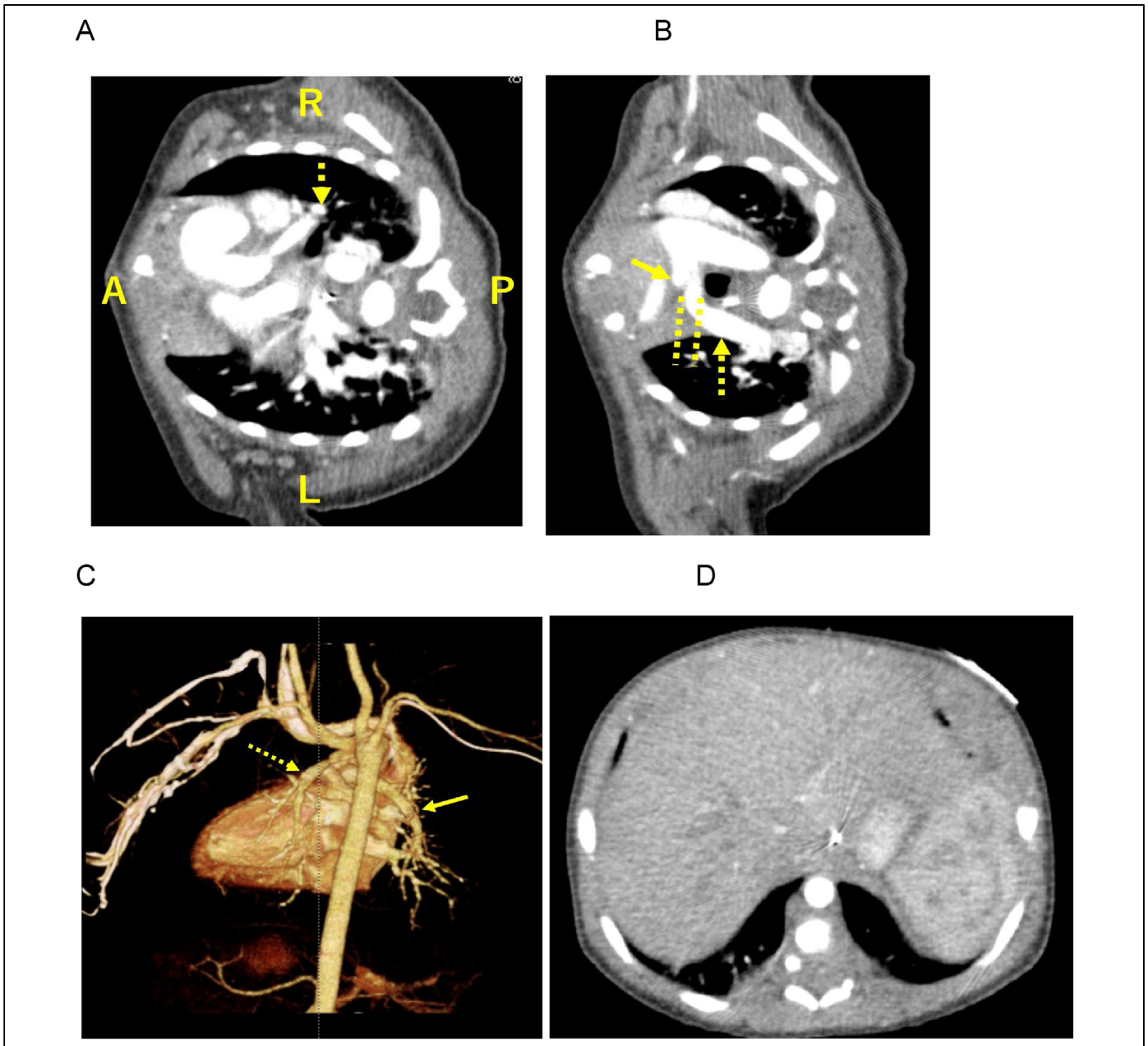

Fig. 2. Postnatal thoracic contrast-enhanced computed tomography (CT). A and B were inverted for comparison with the echocardiography image. (A) The dotted arrow indicates the right pulmonary artery. (B) The single arrow indicates the brachiocephalic artery, double-dotted line indicates the left subclavian artery, and dotted arrow indicates the left pulmonary artery. (C) Three-dimensional CT (posterior) view: the single arrow indicates the right pulmonary artery, and the dotted arrow indicates the left pulmonary artery. (D) The spleen was multi-lobulated, which confirmed the diagnosis of left isomerism.

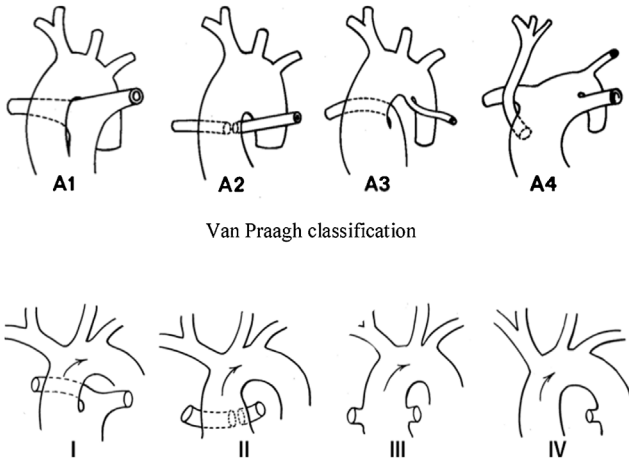

Fig. 3. Van Praagh classification and Collett and Edwards classification [2,3].

when the patient was 2 months old and weighed 4.2 kg. The right pulmonary artery was dissected from the common arterial trunk, and the peripheral end of the left ductus arteriosus was dissected from the left pulmonary artery. The right ventricular outflow tract was reconstructed using a three-valve pericardial roll made from bovine pericardium, and the ventricular septal defect was closed. The patient was discharged without any complications at 35 days after surgery.

Discussion

The C–E classification of TrA was reported in 1949 and the VP classification in 1965; VP type A3 does not exist in the C–E classification (Fig. 3) [1–3]. According to Calder et al., VP type A3 comprises 8% of all TrA cases, and cases such as the present one, with an additional aortic arch and a contralateral ductus arteriosus, are extremely rare and occur in only 2% of the patients [3]. In addition, narrowing or closure of the ductus arteriosus in VP type A3 can cause hypoplasia or closure of the unilateral pulmonary artery [4,5]. Therefore, an accurate diagnosis of this rare type of TrA should be made prenatally, or in a timely fashion postnatally to plan treatment immediately after birth.

In the present case, when the fetus' horizontal planes were scanned, the origin of the left pulmonary artery was not clearly visible on two-dimensional fetal echocardiography. However, on color Doppler images, the right pulmonary artery originating from the common arterial trunk and the left pulmonary artery originating from the brachiocephalic artery were observed almost simultaneously while only slightly moving the probe.

The coalescence of colors of the right pulmonary artery and the brachiocephalic artery led to a misperceived appearance of bifurcation of the bilateral pulmonary arteries only according to the C–E classification. Although each factor should have been carefully investigated from an anatomical and physiological standpoint, additionally considering the VP classification from the beginning might have allowed us to doubt the diagnosis of the A3 type and focus on detecting the origin of the left pulmonary artery and ductus arteriosus more carefully. The sagittal view can be useful to detect ductus arteriosus since the flow observed in the vessel heads toward the caudal direction around the aortic arch, which is usually not observed in a healthy fetus or other types of TrA, except in type A3.

Usually, intracardiac repair open-heart surgery is performed during the neonatal period in TrA; however, it is associated with a high mortality rate of about 10% [6]. Therefore, a temporary procedure is performed at times to avoid open-heart surgery in the neonatal period, and a staged definitive intracardiac repair is performed later [7]. The present case required central pulmonary artery reconstruction; hence, a two-stage intracardiac repair was planned. VP type A3 requires maintenance of patency of the ductus arteriosus and unilateral pulmonary artery banding. In addition, while waiting for the intracardiac repair after the temporary procedure, we avoided introducing major changes in bilateral pulmonary vascular resistance. To avoid high pulmonary blood flow only to the left lung on the side of the ductus arteriosus, the dose of PGE_1 was adjusted to maintain similar blood flow velocities at the left ductus arteriosus and right pulmonary artery banding site. Postoperative cardiac catheterization showed mild stenosis of the left pulmonary artery, but no unilateral pulmonary hypertension. As described above, the two-stage treatment for TrA type A3 requires careful control of the patency of the ductus arteriosus using PGE1 and a longer hospital stay. However, this technique can be considered as one of the efficient strategies to achieve better results of total repair for TrA VP type A3, as it allows us to wait as the patient grows and matures.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at doi:https://doi.org/10.1016/j.jccase.2019.03.009.

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