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CASE REPORT

Prenatal diagnosis of the rare association of common arterial trunk and double aortic arch

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Key Clinical Message

Common arterial trunk with associated double aortic arch is a very rare constellation of congenital heart disease. Prenatal diagnosis allows for surgical repair prior to development of respiratory morbidity, which is otherwise described in all cases with this association.

Keywords

Common arterial trunk, double aortic arch, prenatal, vascular ring.

What is already known about this topic?

- Common arterial trunk and double aortic arch comprise a rare association of congenital cardiovascular disease.
- Surgical correction of common arterial trunk (with or without associated abnormalities) is recommended in the neonatal period.
- All prior case reports of the association of common arterial trunk and double aortic arch describe postnatal respiratory morbidity.

What does this study add?

- We report the first prenatal diagnosis of the association of common arterial trunk and double aortic arch.
- Respiratory morbidity may be avoided in the newborn with early diagnosis and surgical intervention.

Introduction

A 36-year-old gravida 2, para 0 woman presented for fetal echocardiography at 20 5/7 weeks gestation due to abnormal cardiac views on routine obstetric scan. The family history was noncontributory and maternal cell-free DNA testing was negative. Common arterial trunk, aortic dominant type [1], with a right aortic arch was identified. A Level II obstetric ultrasound, genetics consultation, and microarray chromosomal analysis were recommended. No further anatomic abnormalities were identified. Amniotic microarray analysis was performed, showing a 46,XY,del 22q11 karyotype. A follow- up fetal echocardiogram at 25 2/7 weeks gestation was performed for serial assessment of truncal valve function. At that time, a right-dominant double aortic arch was identified on a high axial 3-vessel tracheal view. The double aortic arch was most clearly seen with color Doppler imaging (Fig. 1). No further anatomic abnormalities were identified on serial echocardiograms prior to delivery.

The remainder of the pregnancy was unremarkable. At 38 5/7 weeks, a 2.6 kg male infant was born vaginally. Transthoracic echocardiography and computed tomography (CT) angiography of the chest on the first day of life confirmed the diagnosis of common arterial trunk with double aortic arch (right arch dominant) (Fig. 2). The CT also suggested tracheal compression at the level of the arches.

The patient did not require preoperative respiratory support. On day of life 4, the infant underwent complete open repair including pericardial patch closure of the ventricular septal defect, placement of an 8-mm

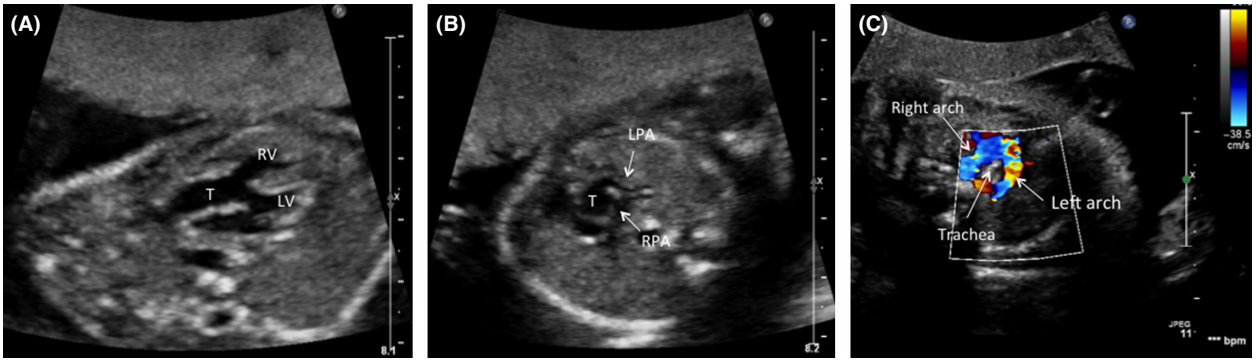

Figure 1. Fetal echocardiogram. (A) Transverse view at the outflow tract level; (B) Short-axis view of common arterial trunk and branch pulmonary arteries; (C) A 3-vessel tracheal view of the double aortic arch with color Doppler. RV, right ventricle; LV, left ventricle; T, common arterial trunk; LPA, left pulmonary artery; RPA, right pulmonary artery.

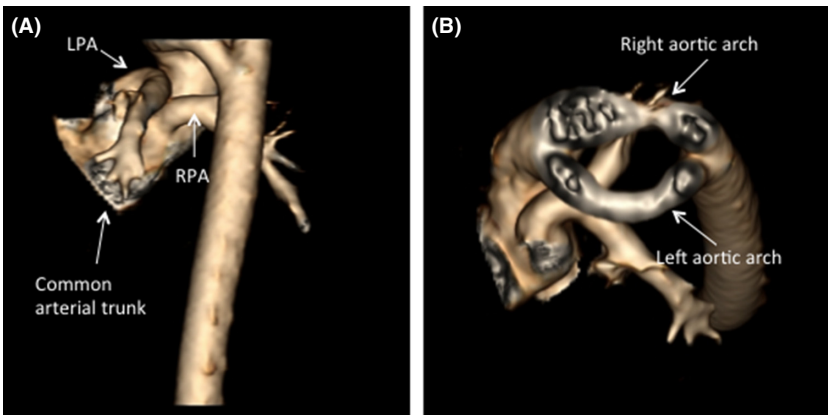

Figure 2. CTA-derived three-dimensional reconstruction images. (A) posterior view of common arterial trunk; (B) cranial view of right and left aortic arches forming complete vascular ring. LPA, left pulmonary artery; RPA, right pulmonary artery.

pulmonary valve homograft from the right ventricle to pulmonary artery, and division and oversewing of the left aortic arch. Postoperative bronchoscopy showed no tracheal compression. The postoperative course was uneventful and the infant was discharged on postoperative day 15.

We report the 10th known case of common arterial trunk with double aortic arch, and the first in utero diagnosis in the literature [2–7]. Other reported cases have presented with respiratory symptoms of a vascular ring, which most commonly include respiratory distress and even respiratory arrest in infants. In contrast, our patient did not experience preoperative respiratory distress, suggesting that prenatal diagnosis may have reduced our patient's morbidity by allowing planning for airway management, identification of a contraindication to transesophageal echocardiography, and early surgical repair of the double aortic arch at the time of common arterial trunk repair. The complete prenatal anatomic diagnosis and identification of 22q11 deletion assisted greatly in prenatal parental counseling and discussion of expectations for the infant's postnatal course.

Common arterial trunk with double aortic arch remains a very rare condition. There is a clear association of DiGeorge syndrome with both conotruncal defects and aortic arch anomalies, leading to the recommendation for genetic testing at the time of common arterial trunk identification on the initial prenatal echocardiogram. While our patient does have a documented 22q11 deletion, not all cases have had genetic or clinical diagnoses of DiGeorge syndrome. Current recommendations suggest that microarray testing should be done for patients with conotruncal and arch anomalies [8].

This case also highlights the importance of serial examination when an arch abnormality is diagnosed. As the majority of double aortic arches have a dominant arch, the minor arch may be difficult to identify early in

gestation and only be visible later in gestation. On repeat fetal echocardiographic scanning in the 3-vessel tracheal view, a small left-sided aortic arch was suspected on two-dimensional imaging and this was in fact confirmed on color Doppler imaging at 25 weeks. The identification of three arches (dominant aortic arch, minor aortic arch, and ductal arch) in the 3-vessel view is diagnostic of a double aortic arch [9].

Conflict of Interest

None declared.

References

1. Russell, H., M. Jacobs, R. Anderson, C. Mavroudis, D. Spicer, E. Corcrain, et al. 2011. A simplified categorization for common arterial trunk. J. Thorac. Cardiovasc. Surg. 141:645–653.
2. Alboliras, E., S. Lombardo, and J. Antillon. 1995. Truncus arteriosus with double aortic arch: two-dimensional and color flow doppler echocardiographic diagnosis. Am. Heart J. 129:415–417.
3. Bhan, A., M. Gupta, M. J. S. Kumar, S. Khotari, and G. Gulati. 2006. Persistent truncus arteriosus with double aortic arch. Pediatr. Cardiol. 27:378–380.
4. Huang, S., C. Wang, W. Su, J. Chu, and M. Hwang. 2008. The rare association of truncus arteriosus with a cervical double aortic arch presenting with left main bronchial compression. Cardiology 111:16–20.
5. Imai, K., K. Tsukuda, H. Sakazaki, and K. Fujiwara. 2013. Persistent truncus arteriosus with double aortic arch and mitral stenosis. Pediatr. Cardiol. 34:2024–2026.
6. Pacilio, G., G. Palma, M. Russo, C. Vosa, and R. Calabro. 1991. Truncus arteriosus and double aortic arch associated with DiGeorge syndrome. Tex. Heart Inst. J. 18:206–208.
7. Paul, J. F., and A. Serraf. 2002. Truncus arteriosus and double aortic arch. Circulation 105:e170.
8. Donofrio, M. T., A. J. Moon-Grady, L. K. Hornberger, J. Copel, M. Sklansky, A. Abuhamad, et al. 2014. Diagnosis and treatment of fetal cardiac disease: a scientific statement from the American Heart Association. Circulation 129:2183–2242.
9. Trubo, D., C. Bravo, T. Alvarez, R. Perez, F. Gamez, and J. De Leon-Luis. 2015. Prenatal sonographic features of a double aortic arch. J. Ultrasound Med. 34:1921–1927.