

Prenatal diagnosis & postnatal outcome of Right Aortic Arch - A 10-year cohort study

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INTRODUCTION

Right-sided aortic arch(RAA) is an aortic arch abnormality with an incidence of 1 in 1000 It can either be an isolated finding or be a part of a complex cardiac malformation.

The diagnosis of right-sided aortic arch is important in order to identify possible associated cardiac and non-cardiac defects and chromosomal abnormalities, especially 22q11 microdeletion and trisomy 21

OBJECTIVE

In our study, we aimed to review the fetuses diagnosed with RAA, evaluate the associated anomalies, and study their outcomes.

DESIGN

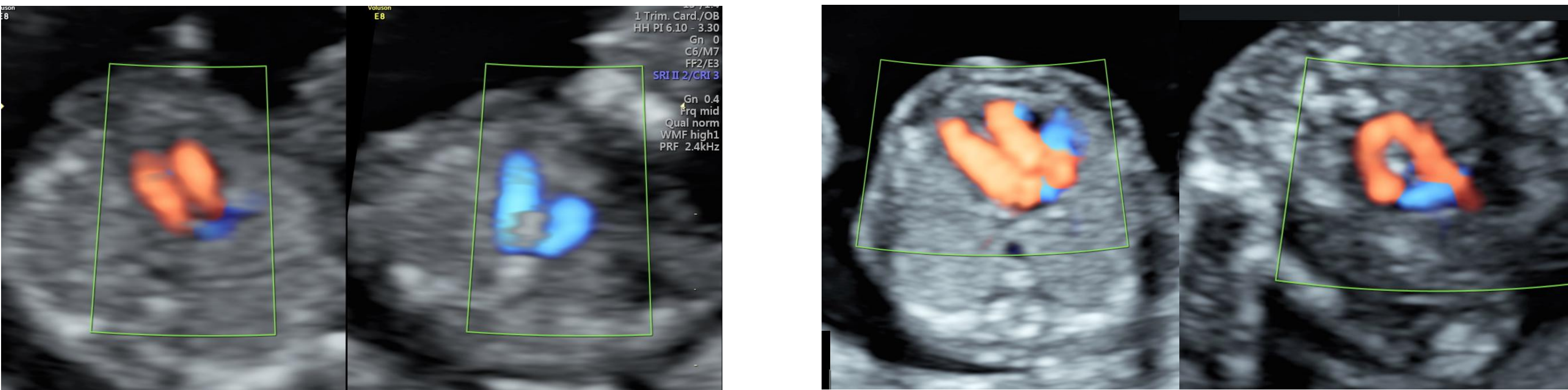
A retrospective 10-year cohort including all the fetuses with RAA that were prenatally diagnosed in our Maternal & Fetal Medicine referral center.

METHODOLOGY

A right-sided aortic arch was diagnosed when the transverse arch was to the right of the trachea in the three-vessel-trachea view. Prenatal echocardiography was performed by fetal medicine specialists and postnatal echocardiography was done by a pediatric cardiologist. Chromosomal Microarray for diagnosis of 22q11 deletion was offered to each patient. The data was collected retrospectively from the electronic medical records. The postnatal outcome was obtained by personal interrogation and reviewing the baby’s medical records.

RESULTS

- There were a total of 17 fetuses with Right aortic arch during the study period January 1, 2013 to January 1, 2023.
- 6 fetuses (35.3%) had isolated RAA
- 11 fetuses (64.7%) had other associated anomalies.
- The most common associated cardiac anomaly was Tetralogy of Fallot (44%).
- Extracardiac malformations were observed in 6 fetuses (35.2%).
- Though genetic testing with Chromosomal microarray was offered to all, only 5 patients (29.4%)went ahead for genetic study, while others declined.
- There was one fetus with Trisomy 21 and one with 22q11 deletion.
- Those with associated anomalies went for medical termination of pregnancy whereas those with isolated RAA continued pregnancy
- Postnatally, the babies with isolated RAA were all asymptomatic except 1 baby with respiratory symptoms.
- None of them had significant tracheal compression or required surgery



1st trimester- Four chamber & Right aortic arch 2nd trimester- Four chamber & Right aortic arch

DISCUSSION

- A right aortic arch occurs when the aortic arch traverses over the right bronchus instead of the left bronchus. Embryologically, this develops when the right dorsal aorta persists and the left dorsal aorta regresses.
- Depending on the formation of the other branches of the aortic arch, various forms of the right aortic arch can develop. This includes a mirror type right aortic arch, defined by the first branch originating from the aorta being the left innominate artery, the second being the right common carotid artery, and the third being the right subclavian artery.
- Right aortic arch with an aberrant left subclavian artery causes a vascular ring, which can be associated with swallowing difficulties (dysphagia lusoria) in children after they begin to eat solid foods. Rarely, it can be associated with stridor and recurrent croup if there is compression of the trachea

CONCLUSION

- Prenatal diagnosis of RAA is important as it can be associated with other cardiac or non-cardiac abnormalities, 22q11 microdeletion, trisomy 21 or other chromosomal abnormalities.
- Chromosomal Microarray should be offered in any case of RAA, irrespective of co-existing malformations.
- Isolated RAA carries a good prognosis and appropriate counseling should be done to alleviate maternal fears and anxiety regarding the continuation of pregnancy

REFERENCES

- Yerlikaya G, Efetürk T, Springer S, Reischer T. Prenatal detection of right aortic arch. Arch Gynecol Obstet. 2019 Apr;299(4):933-938.