

Prenatal diagnosis of a Right Aortic arch and Right patent ductus arteriosus – not often an innocent combination

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Objectives

The co-existence of a right aortic arch and a right ductus arteriosus although considered rare, has been described to be as high as 10-18% in prenatal series. We present a small case series of three cases with a Right Aortic Arch (RAA) and a Right Ductus arteriosus (rDA) associated with significant congenital heart defects.

Methods

A database search of fetal echocardiograms from 2012-2021 in our institution was performed for the identification of cases with right aortic arch and a right ductus arteriosus (rDA). Gestational age (GA) at diagnosis, prenatal and postnatal clinical presentation, imaging characteristics, genetic associations, as well as prenatal and perinatal management and short-term outcome are reported.

Results

Three prenatal cases of a right aortic arch (RAA) with (rDA) were identified.
Case A: A fetal echocardiogram (echo) performed at 27 weeks gestation (GA) for a two-vessel umbilical cord and intrauterine growth retardation, revealed normal segmental anatomy, suspicion of a small ventricular septal defect (VSD), a RAA and an rDA. Patient A was born at 37 weeks GA and postnatal echo revealed a moderate sized VSD, a RAA, a rDA and mild stenosis of the right pulmonary artery. Due to congestive heart failure symptoms underwent successful VSD closure at 6 weeks of life and is doing well at 7 months of age. Microarray was non-diagnostic. Patient Case B: A fetal echo was performed at 30 weeks GA for maternal diabetes revealing normal segmental cardiac anatomy, right ventricular (RV) dominance and a RAA with an rDA with suboptimal imaging of the branch pulmonary arteries (PAs) and the aortic isthmus. Patient was born at 37 weeks GA and postnatal echo revealed a RAA, bilateral DA with a ductal dependent, hypoplastic left pulmonary artery stenosis and possible coarctation of the aorta. He was operated on at 12 days of life and underwent bilateral PDA ligation and LPA plasty and is alive and well at 9 months of age. Microarray was not diagnostic. Case C: A fetal echo was performed at 26 weeks of age for a possible VSD and pulmonary stenosis. The diagnosis of Tetralogy of Fallot with a right aortic arch and a small rDA was made. Patient was born prematurely at 36 and 2/7 weeks GA and the diagnosis was confirmed by postnatal echocardiogram. No intervention was performed as of yet and the patient is stable at 4 weeks of age awaiting surgery. Microarray is pending.

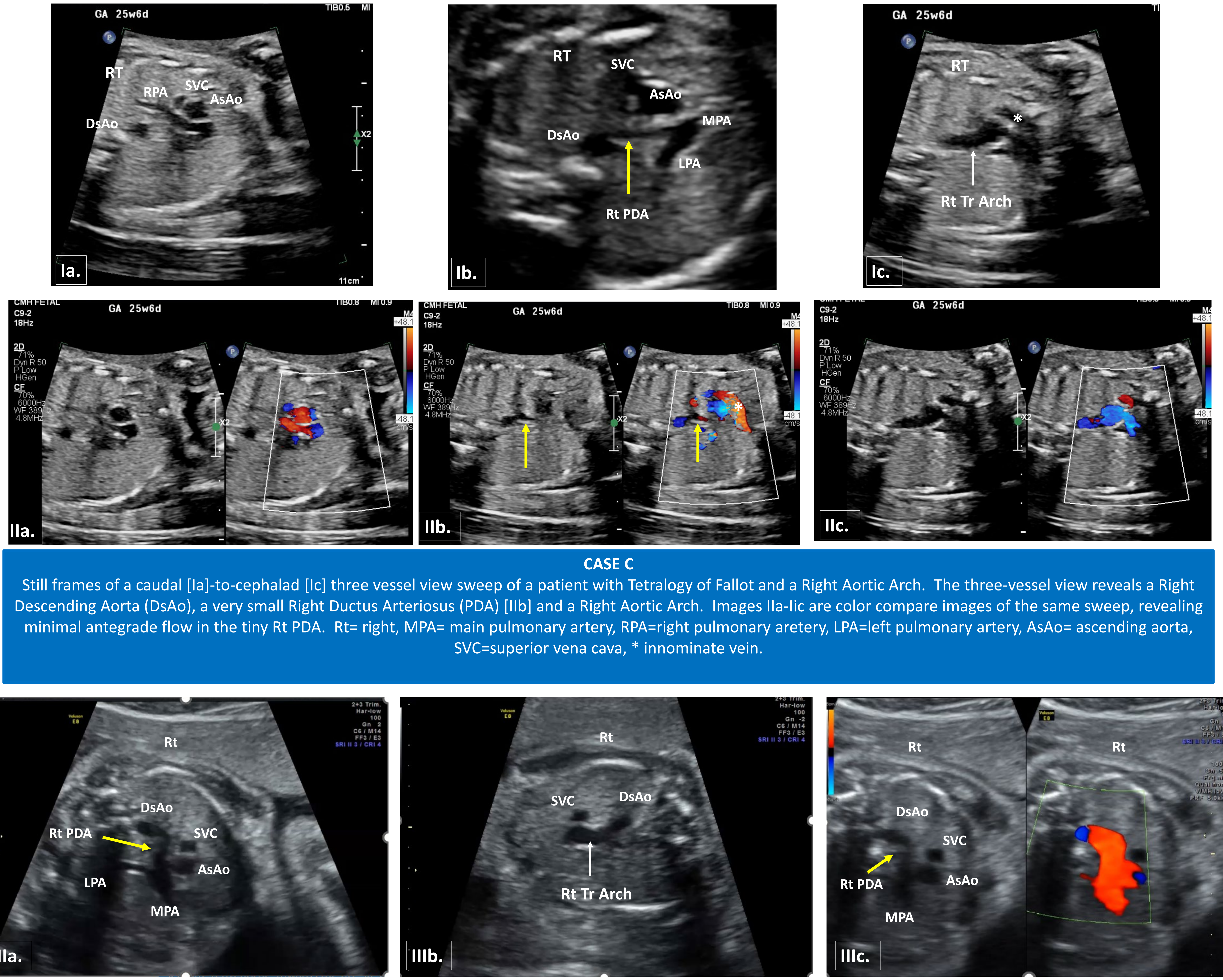
Conclusion

The combination of a right aortic arch with a rDA although uncommon, can frequently be associated with significant congenital heart disease. Prenatally, sweep of the 3 vessel-tracheal (3VT) view in the upper fetal mediastinum and the sagittal arch views (aortic and ductal) are pivotal for the evaluation of the branch PAs, aortic arch and DA situs and can assist in the diagnosis. In RAA and rDA this view is abnormal and the abnormality should be recognized, since the classic V-sign is on the right of the trachea. Evaluation of intracardiac anatomy is imperative since it may reveal coexisting defects. Bilateral DAs warrant evaluation of the PAs to exclude their ductal dependency. Postanatal imaging studies such as echocardiogram and Computed Tomographic Angiography will elucidate further details of the great vessel anatomy.

References

1. Ekiz A, Gul A, Uludogan M, Bornaun H. Prenatal diagnosis and postnatal outcome of persistent right ductus arteriosus: a report of three cases. J Med Ultrasonics 2015;42:571-574
2. Chiappa E, Ridolfi C, Cordisco A. The multiform sonographic spectrum of arterial duct in right aortic arch. The International Journal of Cardiovascular Imaging.

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CASE C
Still frames of a caudal [Ia]-to-cephalad [Ic] three vessel view sweep of a patient with Tetralogy of Fallot and a Right Aortic Arch. The three-vessel view reveals a Right Descending Aorta (DsAo), a very small Right Ductus Arteriosus (PDA) [I Ib] and a Right Aortic Arch. Images IIa-IIIc are color compare images of the same sweep, revealing minimal antegrade flow in the tiny Rt PDA. Rt= right, MPA= main pulmonary artery, RPA=right pulmonary aretery, LPA=left pulmonary artery, AsAo= ascending aorta, SVC=superior vena cava, * innominate vein.

CASE A
Still frames of a caudal [IIIa]-to-cephalad [IIIb] sweep of the three vessel view in the patient with Rt Aortic Arch and Ventricular Septal Defect. The sweep reveals a Right Descending Aorta (DsAo), a Right Ductus Arteriosus (PDA) and a Right Aortic Arch. Image IIIc, is a color compare image showing same flow direction in both Ductal and Transverse Aortic Arch (Tr Arch). Rt= right, MPA= main pulmonary artery, LPA=left pulmonary artery, AsAo= ascending aorta, SVC=superior vena cava.