Prenatal Delineation of Coronary Anatomy in D-transposition of Great Arteries

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Abstract

Dextro-transposition of the great arteries (D-TGA) is the second most common cyanotic congenital heart disease with variable coronary artery anatomy. Outcomes have improved significantly with the advent of the arterial switch procedure for this defect, with coronary artery anatomy being a very important parameter for both short and long term outcomes following surgical repair. Assessment of coronary artery anatomy is usually undertaken in the postnatal period by transthoracic echocardiography. Prenatal delineation will help with surgical planning, parental counseling and tertiary care referral as needed due to the critical importance of this variable in successful outcome. We describe our experience with prenatal coronary artery assessment in a cohort of patients with fetal diagnosis of D-TGA and highlight the importance of its assessment with both 2-dimensional (2D) and Color Doppler imaging, especially after 25 weeks gestation.

Introduction

The prevalence of major congenital heart disease at birth is close to 1%[1,2,3]. Dextro-transposition of the great arteries (D-TGA) is the second most common cyanotic congenital heart defect, accounting for 5% of all congenital heart defects[4]. D-TGA has a prevalence of 20 to 30 per 100,000 live births, with a 2:1 male predominance (particularly large-term males).[4] In D-TGA, the aorta arises from the right ventricle with the pulmonary artery arising from the left ventricle resulting in ventriculoarterial discordance and parallel circulations. Despite advances in fetal echocardiography, the prenatal diagnosis of D-TGA remains low in various series[16]. Arterial switch operation (ASO) has become the preferred technique for anatomic correction of D-TGA, offering the advantage of a systemic left ventricle over the Mustard/Senning (atrial switch) procedure which results in a systemic right ventricle. Delineation of coronary artery anatomy in D-TGA is essential prior to surgical planning for the arterial switch procedure since certain types of coronary anatomy necessitates a modification of the surgical transfer technique. We sought to review our experience with the prenatal definition of coronary artery anatomy in a cohort of fetuses with D-TGA confirmed on postnatal echocardiogram and intraoperatively.

Methods

A retrospective review of fetal echocardiograms with prenatal diagnosis of D-TGA at Akron Children's Hospital and Cleveland Clinic Children's Hospital over a 10 year period from 2008 to 2018 was performed. Studies were identified from review of fetal echocardiographic database and chart review. Demographic variables, including maternal age, and gestational age at diagnosis were recorded. Fetal echocardiographic parameters including presence/absence of ventricular septal defect (VSD) and gestational age (GA) at delineation of coronary artery anatomy were recorded. Postnatal echocardiogram with regards to coronary artery anatomy, as well as surgical confirmation of coronary artery anatomy (at the time of the arterial switch procedure) was recorded. Studies were performed using a Siemens S 3000 machine using the 8 MHz curved array transducer with harmonic imaging and Phillips EPIQ machine. 2-dimensional (2D) as well as Color Doppler Imaging was utilized for the assessment of cardiac anatomy, including coronary artery anatomy. Color Doppler velocity scale was adjusted between 30 and 45 cm/s to image flow through the coronary arteries. The echocardiographic images were individually reviewed to assess for the documentation and confirmation of the coronary artery patterns.

Results

We identified a total of 34 fetuses with a prenatal diagnosis of D-TGA over the 10 year period of the study. In 14/34 fetuses, partial or complete delineation of coronary artery anatomy was attempted (Table 1). The average age of patients was 28 years (range 19 to 41 years). Average GA at diagnosis was 28 weeks (range 23 to 31 weeks). 11/14 patients had delineation of coronary artery anatomy at or more than 25 weeks GA. 12/14 fetuses had D-TGA with intact ventricular septum (IVS) and 2/14 patients had D-TGA with VSD. In 11 of 14 fetuses, complete delineation of coronary artery anatomy could be done prenatally, with abnormal anatomy in 2 fetuses. 9/14 fetuses had usual coronary anatomy for D-TGA (Figures 1 and 2). In 3/14 cases, the right coronary artery system could not be well delineated prenatally. 2/14 fetuses had a circumflex coronary artery arising from the right coronary artery (Figure 3). A coronary artery arising from the right coronary cusp and traversing in the left AV groove was a marker for anomalous origin of the circumflex from the right coronary artery (Figure 2). Coronary anatomy as well as the cardiac anatomy diagnosed prenatally was confirmed postnatally on the transthoracic echocardiograms and intraoperatively during the arterial switch procedure.

Discussion

Anomalies of coronary arteries occur in both the origin and distribution, with further anomalies secondary to intramural course in D-TGA[14]. Presence of single coronary artery, anomalous circumflex coronary artery or intramural course may complicate the surgical technique of the arterial switch procedure in this condition[15]. The presence of a ventricular septal defect or of side-by-side great vessels should alert the imager/cardiologist to an increased likelihood of coronary anomalies. In almost all patients, the coronary arteries arise from the facing sinuses. Delineation of coronary anatomy is crucial in the pre-operative assessment and surgical management of these patients. In approximately 60% of patients, the coronary arteries arise from their appropriate sinuses and branch normally, a pattern most often seen with the aorta anterior and to the right of the pulmonary artery. The coronary arteries usually take the shortest course to reach their distribution, and because the aorta is anterior, the left main and circumflex coronary arteries pass anterior and leftward to the right ventricular outflow tract. This usual pattern of coronary arteries was found in 12/14 patients in our study (85.7%). The next most common variant is origin of the left circumflex coronary artery from the right coronary artery and coursing posterior to the pulmonary artery, which is seen in about 20% of patients, usually with side-by-side great vessels. This was seen in 2/14 patients (14.3%) in our study. The coronary arteries may be completely inverted, with the right coronary artery arising from a left anterior sinus and the left coronary artery arising from the right posterior sinus. Finally single coronary artery is a rare variant as well as intramural and inter-arterial course, which were not found in our study, given the small sample size and associated rarity.

ASO has become the procedure of choice for surgical repair of D-TGA[5-8]. Compared with atrial-level Mustard and Senning repairs, correction at the arterial level restores the left ventricle as the systemic pumping chamber and is associated with improved long term outcomes [9-11]. Accurate identification of coronary artery anatomy is extremely important for the success of the ASO, therefore much attention has been paid to the relationship between the coronary pattern and outcome [12-14]. Prenatal detection of D-TGA used to be rare, but is now becoming more common due to advances in prenatal ultrasound screening [16]. Although some cases are missed during the prenatal period, with the newer AIUM guidelines to assess the outflow tracts on prenatal ultrasound, improved referral and detection is anticipated.

In current practice, the coronary artery anatomy is primarily assessed after birth, but we propose that prenatal delineation of coronary artery anatomy can be performed with careful attention to imaging details. However, we acknowledge that various factors such as fetal position, gestational age and maternal body habitus as well as multiple gestation could influence the successful assessment of cardiac and coronary artery anatomy. In our study, we were able to assess coronary artery anatomy in 41% of patients, with improved chances of success after 25 weeks gestation. Prenatal coronary artery assessment appears to provide multiple benefits, especially since ASO have increasingly been performed sooner after birth [17-20]. It also aids in the planning of the postnatal course and in counselling the parents about the potential variations in surgical technique if they were found to be abnormal. Additionally, patients can be referred to surgical centers of excellence for the more complex variants to ensure successful outcomes as needed. The coronary artery anatomy in fetuses with D-TGA has not been widely described, except for one study by Takashi et al[21] which reported 3 cases of prenatal identification confirmed at angiography at the time of balloon atrial septostomy postnatally. Our study has the largest number of patients with prenatally described coronary artery anatomy with the help of both 2D and Color Doppler imaging. As mentioned in the previous paper, assessment of the semilunar valves in the short axis as well as in the long axis, ideally after 25 weeks gestation, enables identification of coronary arteries. This could be due to the fetus being bigger with better visualization of cardiac anatomy. Careful attention should be given to a coronary artery arising from the right sinus and coursing towards the left atrioventricular groove, since this would raise suspicion for anomalous origin of the circumflex coronary artery. The two patients in our study who had anomalous origin of the circumflex coronary artery were confirmed with postnatal imaging with the surgeon appropriately modifying surgical technique for coronary transfer. Fetal echocardiography additionally provides unique angles of interrogation through the collapsed lungs which may be an advantage compared to the limited acoustic windows from prominent lung artifact on postnatal imaging.

The study by Pasquali et al [22] combined the results of 9 case series in a meta-analysis to estimate the effect of coronary anatomy on mortality after ASO, both overall and adjusted for time. They concluded that common coronary variants have undergone ASO without added mortality compared to those with the usual coronary pattern. However, those with intramural or single coronary arteries have significant added mortality that has persisted over time.

Conclusions

We report on a large series of patients with D-TGA diagnosed prenatally in whom successful identification of coronary artery anatomy was made possible with the help of gray-scale and Color Doppler imaging. This has prognostic significance and aids in prenatal counselling as well as surgical planning. We anticipate that detection rates will only continually improve with newer advances in technology.

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Table 1: Demographic characteristics

Number	Maternal Age (years)	Gestational Age (GA) at Diagnosis (weeks)	Cardiac Diagnosis	Coronary Pattern
1	22	26	D-TGA/IVS	LCA system seen
2	22	24	D-TGA/IVS	LCA system seen
3	19	30	D-TGA/IVS	Usual coronary patter
4	34	28	D-TGA/IVS	Usual coronary patter
5	20	32	D-TGA/VSD	Usual coronary patter
6	29	23	D-TGA/IVS	LCA system seen
7	20	34	D-TGA/VSD	Usual coronary patter
8	26	27	D-TGA/IVS	Usual coronary patter
9	41	30	D-TGA/IVS	Circumflex from RCA

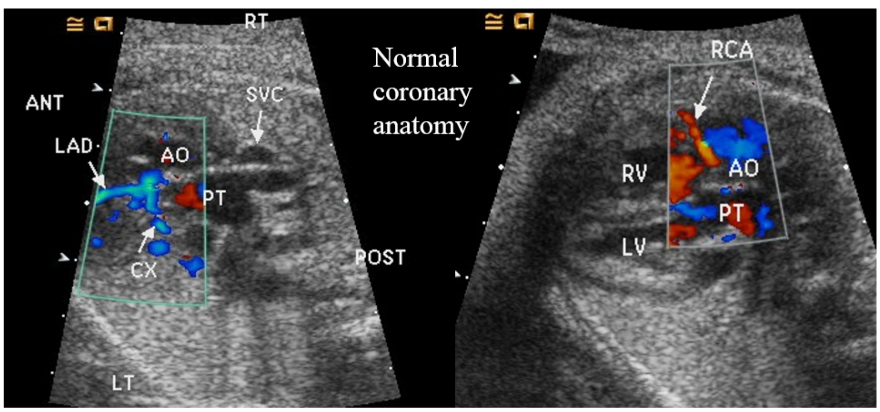
Number	Maternal Age (years)	Gestational Age (GA) at Diagnosis (weeks)	Cardiac Diagnosis	Coronary Pattern
10	33	25	D-TGA/IVS	Circumflex from RCA
11	36	24	D-TGA/IVS	Usual coronary patter
12	20	31	D-TGA/IVS	Usual coronary patter
13	39	29	D-TGA/IVS	Usual coronary patter
14	28	31	D-TGA/IVS	Usual coronary patter

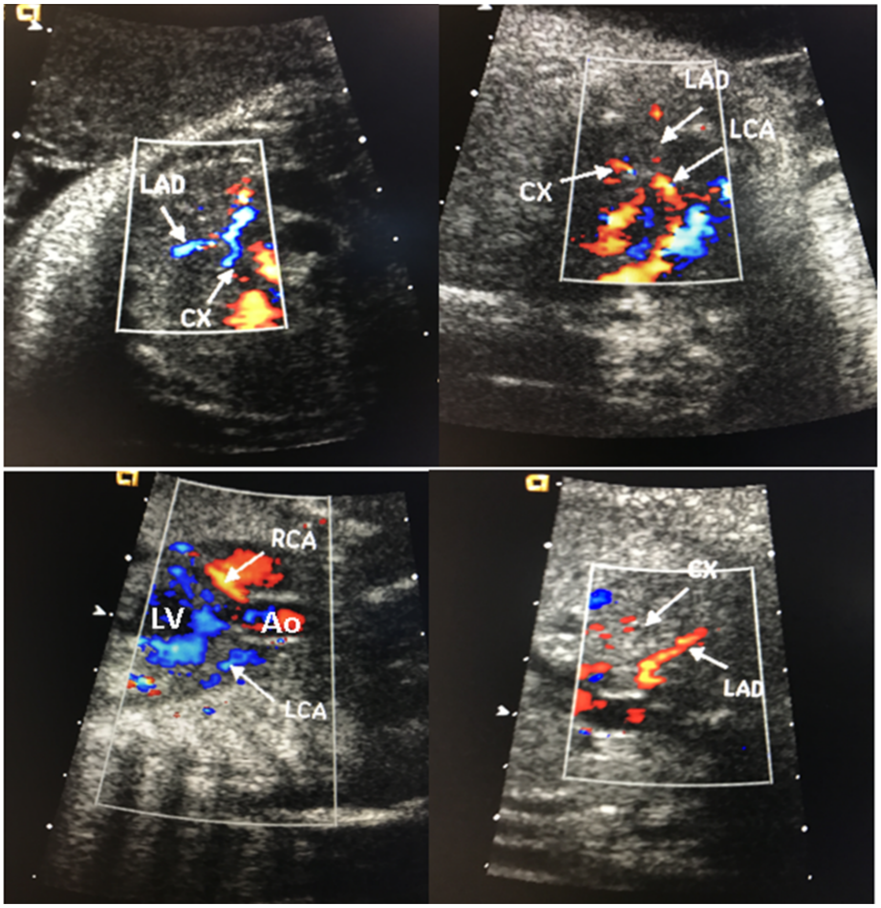
Figure Legends

Figure 1: Color Doppler image showing usual coronary pattern: The left coronary artery arises from the left posterior facing sinus and bifurcates into the left anterior descending and left circumflex coronary arteries and right coronary artery arises from the right posterior facing sinus, coursing in the right AV groove.

Figure 2: Color Doppler image showing usual coronary pattern: The aorta is the anterior vessel. The left coronary artery arises from the left posterior facing sinus and bifurcates into the left anterior descending and left circumflex coronary arteries and right coronary artery arises from the right posterior facing sinus, coursing in the right AV groove.

Figure 3: 2D images: Anomalous origin of the circumflex coronary artery from the right facing sinus and coursing towards the left AV groove.

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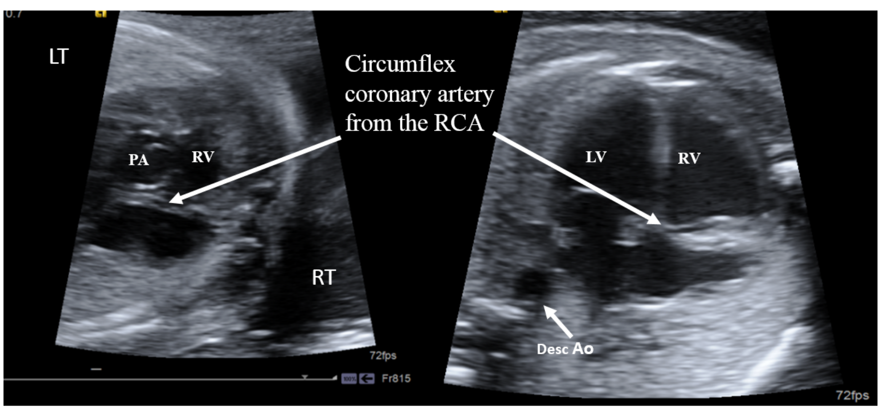
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