Incremental value of fetal spatiotemporal image correlation echocardiography in the diagnosis of tetralogy of Fallot with disconnected pulmonary arteries with ductus arteriosus supplying the left pulmonary artery

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ABSTRACT

Disconnected branch pulmonary arteries are sparsely reported cases in prenatal diagnosis literature. We report a case of tetralogy of Fallot with disconnected branch pulmonary arteries, the left pulmonary artery (LPA) arising from an indirect ductus arteriosus (DA) from the base of the innominate artery in a right aortic arch, diagnosed by fetal echocardiography with 3D/4D spatiotemporal image correlation (STIC) imaging. Prenatal diagnosis led to early neonatal intervention to maintain blood flow to the LPA by stenting of the DA. Fetal echocardiographic evaluation (Voluson E10 systems, GE Healthcare, Zipf) with acquisition of images and volumes in the right ventricular outflow tract and three-vessel trachea view with rendering of 3D/4D STIC volume datasets to display images in high-definition color format. Prenatal evaluation was initially done at 17-week gestation in a 28-year-old pregnant female which showed tetralogy of Fallot (TOF). Subsequent evaluation at 34 weeks with 3D/4D STIC datasets showed a small main pulmonary artery (MPA) continuing into an adequately sized right pulmonary artery. The LPA was very small (Z-score −2.63), with no visible connection to MPA. Rendering of the 3D/4D STIC datasets revealed disconnected pulmonary arteries with the vertical DA from the base of the innominate artery in a right aortic arch, continuing as the LPA. Findings were confirmed on postnatal high-resolution CT pulmonary angiography and cardiac catheterization with subsequent stenting of the ductus. This report highlights the incremental benefit of advanced 3D/4D STIC rendering in accurate prenatal diagnosis of a rare anomaly of TOF with disconnected pulmonary arteries, leading to early neonatal intervention to preserve the blood supply to the left lung.

Keywords: Disconnected branch pulmonary arteries, prenatal diagnosis, spatiotemporal image correlation imaging

INTRODUCTION

Disconnected branch pulmonary arteries originate from an embryological defect in the proximal 6th aortic arch and encompass a spectrum of anomalous origin to the unilateral absence of a branch pulmonary artery.[1] Disconnected LPA, originating from the base of

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the innominate artery, diagnosed antenatally, has been reported previously.[2] We report a case of tetralogy of Fallot with disconnected branch pulmonary arteries with the vertical ductus arteriosus (DA) arising from the undersurface of right-sided aortic arch at the base of the innominate artery continuing as the LPA, diagnosed antenatally with the help of spatiotemporal image correlation (STIC) imaging followed by a successful neonatal ductal stenting.

CASE REPORT

A 28-year-old 4th gravida was referred to us at 17 weeks of gestation with the suspicion of congenital heart disease. Fetal echocardiographic evaluation (Voluson E10 systems, GE Healthcare, Zipf) showed a large malaligned ventricular septal defect with more than 50% aortic override with a small pulmonary artery in the three-vessel view suggesting pulmonic stenosis. There were no associated extracardiac anomalies in the anomaly scan.

Subsequent evaluation at 34 weeks confirmed the presence of a large ventricular septal defect with aortic override [Figure 1a] with a small main pulmonary artery (MPA) (Z-score –5.3) in three-vessel view [Figure 1b]. The right ventricular outflow tract views [Figure 1c-f and Video 1] showed the MPA continuing as a good-sized right pulmonary artery (RPA) (Z = –0.17); the left pulmonary artery (LPA) was very small (Z = –2.63) and appeared disconnected from the confluence [Figure 1f]. The aortic arch was right sided. We obtained 3D/4D STIC volume datasets of the aortic arch and DA (RAB6 probe, GE healthcare, Zipf) in the three-vessel tracheal and sagittal views, and these were then rendered using high-definition color mode to display the images. With the STIC rendering, the origin of the DA from the base of the innominate artery and continuing as the LPA was clearly demonstrated [Figure 2a and Video 2]. There was a constriction in the ductus after its origin; the LPA was discontinuous from the MPA. We advised a planned delivery in our center, anticipating the need for a neonatal cardiac intervention to augment pulmonary blood flow.

Postnatal echo-confirmed prenatal findings and the LPA could be seen as a continuation of the DA. A high-resolution 256 slice computed tomography (CT) pulmonary angiography was done to delineate the anatomy and it confirmed the prenatal STIC imaging findings [Figure 2b]. Cardiac catheterization and angiography were undertaken with an intent to stent the DA. Angiography [Figure 3a and Video 3] showed a constricted ductus arising from the right aortic arch and continuing as the LPA. There was an excellent correlation between the prenatal STIC imaging, postnatal CT reconstruction, and angiography imaging [Figure 2a-c]. Ductal stenting was done with 16 mm × 3 mm BioMime stent placed at the junction between the right DA and stenotic origin of LPA under fluoroscopic guidance, thus maintaining the blood flow to the LPA [Figure 3b]. The baby was reviewed after 1 month, and echocardiography

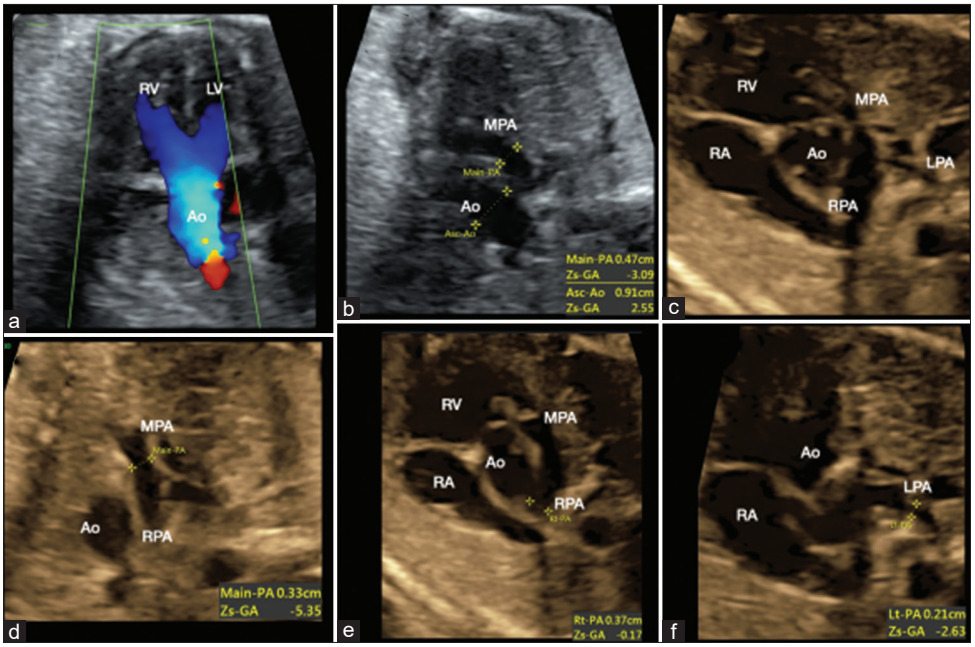

Figure 1: Two-dimensional fetal echocardiography at 34 weeks' gestation. (a) A large over-riding Ao arising from 2 balanced ventricles (LV and RV); (b) is a three-vessel view showing a small pulmonary artery (MPA) *vis-a-vis* a much larger Ao; the respective Z-scores are also shown. (c) Is a RV outflow tract view showing a narrow MPA continuing as the RPA. A very diminutive LPA is also seen. (d and e) The measurement of the size of the main and right pulmonary arteries with Z-scores, respectively. (f) The distal LPA with its measurement and Z-score. RA: Right atrium, RV: Right ventricle, Ao: Aorta, RPA: Right pulmonary artery, LPA: Left pulmonary artery

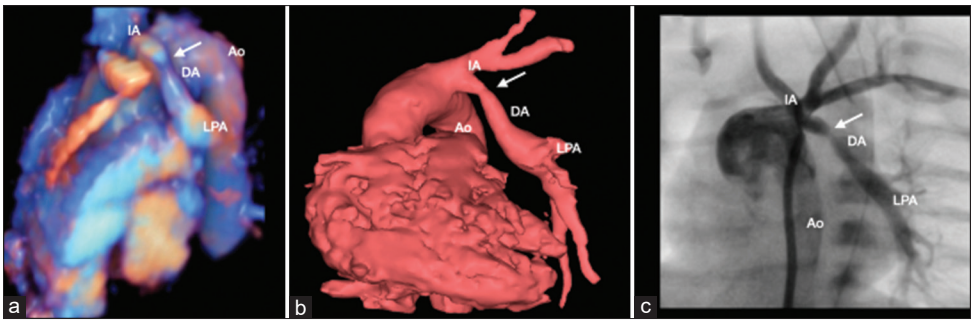

Figure 2: Co-relation of anatomy between prenatal STIC rendering (a), postnatal 256 slice CT based reconstruction (b) and cardiac catheterization angiography (c). There is excellent correlation of the origin of the ductus (DA) from the base of the IA and continuing as the LPA in all 3 images. The constriction (arrow) in the beginning of the ductus is also correlated in all the images. STIC: Spatio-temporal image correlation, LPA: Left pulmonary artery, IA: Innominate artery, DA: Ductus arteriosus

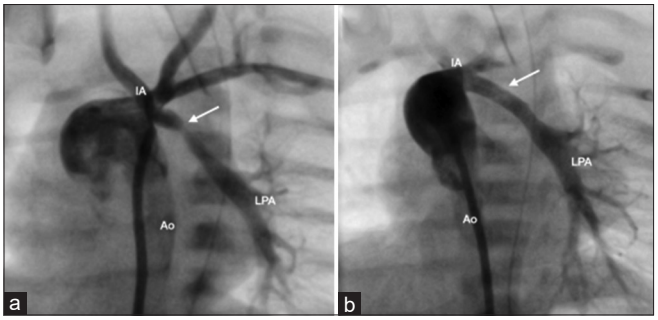

Figure 3: Cardiac catheterization and angiography before (a) and after stenting of the DA (b). The constriction (arrow) in the ductus is eliminated by the procedure. IA: Innominate artery, LPA: Left pulmonary artery, DA: Ductus arteriosus

showed patent stent flow with a good growth of the LPA (Z-score 0.42). The baby is planned for an elective surgical repair at an older age.

DISCUSSION

This case demonstrates the feasibility and incremental benefit offered by new techniques such as 3D/4D STIC imaging in elucidating complex anatomic details during prenatal life, thereby enabling well-planned perinatal management. STIC imaging is a novel ultrasound technology, in which 3D/4D volume datasets of the fetal heart are acquired using specialized ultrasound probes, capturing multiple frames of the area of interest. Then, the images are displayed in a multiplanar format. The images can then be rendered using various display options, such as high-definition color or glass-body mode, to demonstrate various cardiac structures.[3] In this case, the DA was the sole source of blood flow to the left lung and hence strategies to maintain its patency were critical to ensure the optimal growth of the LPA. We have demonstrated the incremental benefit offered by prenatal 3D/4D STIC imaging in similar critical heart defects before and this report further reiterates its utility in planning perinatal care.[4] The correlation of anatomic details between prenatal STIC imaging, postnatal 256-slide CT reconstruction and angiography was quite precise [Figure 2] and this was instrumental in the successful execution of the ductal stenting procedure [Figure 3].

In the normal left arch development, the proximal part of the left sixth aortic arch forms the proximal (extrapulmonary) LPA, while its distal part forms the DA. The right proximal sixth aortic arch forms the proximal (extrapulmonary) RPA, while the distal obliterates early. Early obliteration of the left proximal part causes the intrapulmonary LPA to connect to the embryological dorsal aorta, which is either the underside of the left (ipsilateral) aortic arch or the base of the innominate artery of the right (contralateral) aortic arch, as is seen in our patient. Early identification of disconnected branch pulmonary arteries enables early intervention to preserve the pulmonary blood supply and subsequent definitive repair.[5] Very few antenatally diagnosed cases are reported in literature, and most have been evaluated retrospectively.[2] This report is unique with respect to the novelty of the imaging modalities used for diagnosis as well the meticulous planning of the perinatal care culminating in a successful clinical outcome.

In conclusion, prenatal diagnosis and precise delineation of the anatomy is feasible for CHD with complex pulmonary artery anatomy using fetal echocardiography assisted by newer techniques like 3D/4D STIC rendering enabling better planning of the perinatal care.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Wenxiu L, Yuan Z, Chaoning H, Bin G, Jiang W, Shuang Y. Prenatal diagnosis of anomalous origin of one pulmonary artery branch by two-dimensional echocardiography: Summary of 12 cases. Cardiol Young 2020;30:39-46.
2. Karmegaraj B, Vaidyanathan B. Right aortic arch, bilateral ductus arteriosus, and anomalous origin of left pulmonary artery from innominate artery in a fetus with normal intracardiac anatomy. Echocardiography 2020;37:796-8.
3. Junior E, Tedesco G, Carillho M, Piexoto A, Carvalho F. 4D fetal echocardiography in clinical practice. Donald Sch J Obstet Gynaecol 2015;9:382-96.
4. Vaidyanathan B, Verma R, Kappanayil M, Kumar RK. Prenatal diagnosis of a large left coronary artery to right ventricle fistula with 3-dimensional/4-dimensional spatiotemporal image correlation rendering followed by successful neonatal transcatheter closure. Circ Cardiovasc Imaging 2022;15:e014247.
5. Krasemann T, Dalinghaus M, van Beynum I, Bogers A, van den Berg G. "Absent" unilateral pulmonary arteries aren't absent, but disconnected. Review of the current literature based on 4 cases. Rev Cardiovasc Med 2022;23:46.