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Prenatal Diagnosis of Complex Double-Outlet Right Ventricle Associated with Atrioventricular Canal Defect : A Case Report Case Report

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Abstract*: Double-outlet right ventricle (DORV) is a rare congenital cardiac malformation in which both great arteries arise predominantly from the morphological right ventricle. Its antenatal diagnosis requires careful segmental analysis of the fetal heart and precise assessment of the ventriculoarterial connections. We report the case of a 19-year-old primigravida referred at 38 weeks and 3 days of gestation for delivery planning after antenatal suspicion of complex congenital heart disease. The pregnancy was complicated by severe fetal growth restriction and severe preeclampsia. Antenatal ultrasound showed ventricular septal defect, ventricular imbalance, malposition of the great arteries with suspected origin from the right ventricle, an anterior aorta, a posterior pulmonary artery, and a narrow subpulmonary pathway suggestive of pulmonary stenosis. Cesarean delivery was performed for maternal rescue after persistent severe hypertension despite medical management. Postnatal transthoracic echocardiography confirmed complex DORV associated with atrioventricular canal defect, right ventricular dominance, moderately hypoplastic left ventricle, and valvular and supravalvular pulmonary stenosis. This case underlines the value of prenatal ultrasound in complex conotruncal anomalies and highlights the importance of multidisciplinary perinatal planning and postnatal echocardiographic confirmation.*

Keywords—Double-outlet right ventricle; Prenatal diagnosis; Fetal echocardiography; Atrioventricular canal defect; Pulmonary stenosis; Severe preeclampsia; Fetal growth restriction

INTRODUCTION

Double-outlet right ventricle (DORV) is a rare and heterogeneous congenital cardiac malformation characterized by the origin of both great arteries predominantly from the morphological right ventricle [1,2]. It belongs to the spectrum of conotruncal anomalies and represents less than 1% of congenital heart diseases [1]. DORV is not a single anatomical entity; rather, it includes a broad range of malformations with variable ventriculoarterial connections and frequent associated intracardiac defects such as ventricular septal defect, atrioventricular septal defect, ventricular hypoplasia, and pulmonary outflow tract obstruction [3,4].

Prenatal diagnosis can be challenging because DORV may mimic other conotruncal abnormalities, particularly tetralogy of Fallot, truncus arteriosus, and complex ventricular septal defect [3,4]. However, fetal echocardiography remains essential for identifying abnormal outflow tract anatomy, defining associated lesions, guiding parental counseling, and organizing delivery in a tertiary center with neonatal intensive care and pediatric cardiology support [3].

We report a case of antenatally suspected complex DORV in a term pregnancy complicated by severe fetal growth restriction and severe preeclampsia. Postnatal echocardiography confirmed DORV associated with atrioventricular canal defect, ventricular imbalance, and pulmonary stenosis.

CASE REPORT

A 19-year-old primigravida, with no history of consanguinity, was referred to our tertiary obstetric unit for delivery planning after the antenatal diagnosis of complex fetal congenital heart disease. Her medical history was unremarkable. She had no known hypertension, diabetes mellitus, cardiac disease, renal disease, autoimmune disease, thromboembolic disease, infection during pregnancy, smoking, or toxic exposure. There was no family history of congenital heart disease or congenital malformations.

The pregnancy was dated according to a reliable last menstrual period and was followed in the private sector. The pregnancy course was marked by detection of a fetal cardiac anomaly during the morphology scan.

At 38 weeks and 3 days of gestation, the patient was admitted for obstetric management. On admission, she was conscious and clinically stable. Blood pressure was 150/100 mmHg and remained elevated on repeat measurements. Urine dipstick testing showed significant proteinuria, and the patient reported headache and visual symptoms. Obstetric examination found positive fetal movements, fetal heart rate of 147 beats/min, and uterine height smaller than expected for gestational age. There was no rupture of membranes or vaginal bleeding.

Maternal laboratory evaluation showed no biological criteria of HELLP syndrome. Platelet count, liver enzymes, renal function, coagulation profile, and lactate dehydrogenase

were within acceptable limits. Proteinuria was confirmed by urinary assessment. The overall maternal presentation was consistent with preeclampsia.

A detailed antenatal obstetric ultrasound showed a live singleton male fetus in cephalic presentation, with normal amniotic fluid volume and a homogeneous placenta located laterally on the right side. Fetal biometry showed a biparietal diameter of 91 mm, corresponding to the 24th percentile; head circumference of 323 mm, corresponding to the 32nd percentile; abdominal circumference of 291 mm, below the 1st percentile; and femur length of 62 mm, below the 1st percentile. The estimated fetal weight was 2196 g, below the 1st percentile for 38 weeks of gestation, consistent with severe fetal growth restriction. Doppler assessment showed abnormal uterine artery Doppler findings, whereas umbilical artery Doppler was preserved.

Morphological assessment did not reveal cranial or abdominal anomalies. Both kidneys and the bladder were visualized. The umbilical cord contained two arteries and one vein. The four limbs were visualized, each with three segments.

Fetal cardiac assessment revealed complex congenital heart disease. Sequential sweeping from the four-chamber view demonstrated a ventricular septal defect, which appeared to represent the obligatory drainage pathway of the left ventricle. The great arteries were malposed and appeared to arise from the right ventricle. The aorta was located anteriorly, while the pulmonary artery was posterior to the aorta. The subpulmonary outflow tract was narrow, and the pulmonary artery was smaller than the aorta, suggesting associated pulmonary stenosis (Fig. 1). Overall, the antenatal findings were highly suggestive of a complex conotruncal anomaly within the DORV spectrum and pulmonary stenosis.

Based on the clinical, biological, and ultrasound findings, severe preeclampsia associated with severe fetal growth restriction and complex fetal cyanotic congenital heart disease was retained. The patient was managed in a tertiary care setting with obstetric, anesthetic, neonatal, and pediatric cardiology support.

During hospitalization, the patient developed severe-range blood pressure reaching 160/110 mmHg, associated with persistent neurosensory symptoms including visual disturbances. Antihypertensive therapy was initiated, and magnesium sulfate was administered for seizure prophylaxis. Because of persistent severe hypertension despite medical management, cesarean delivery was indicated for maternal rescue.

A cesarean section was performed under spinal anesthesia. A live male neonate was delivered from clear amniotic fluid. Apgar score was 9 at 1 minute and 10 at 5 minutes. Birth weight was 2600 g. The newborn was initially pink and reactive and was transferred to the neonatal unit for specialized assessment and management.

Postnatal transthoracic echocardiography was performed on the first day of life. Clinically, the neonate was in good general condition, without signs of heart failure. A systolic murmur was present, and peripheral oxygen saturation was 89%. Echocardiography showed situs solitus and levocardia. Systemic and pulmonary venous returns were normal, and the inferior vena cava was not dilated. At the atrial level, a large 6-mm ostium secundum atrial septal defect and a 3-mm ostium primum atrial septal defect were identified.

The cardiac connections showed a right-sided morphological right ventricle and a left-sided morphological left ventricle. A single common atrioventricular valve was present, with mild-to-moderate central regurgitation. The ventricles were unbalanced, with right ventricular dominance and a moderately hypoplastic left ventricle. An inlet ventricular septal defect was present, shunting from the left ventricle to the right ventricle.

Both great arteries arose from the right ventricle, confirming the diagnosis of DORV. The aortic valve was normal. Valvular and supravalvular pulmonary stenosis were present, with a right ventricle-to-pulmonary artery gradient of 50 mmHg. Both pulmonary artery branches measured 4 mm. There was no evidence of aortic coarctation.

The final postnatal diagnosis was complex congenital heart disease associating atrioventricular canal defect, and DORV, with pulmonary stenosis providing relative protection of the pulmonary circulation. The neonate remained clinically stable from a respiratory standpoint, without signs of heart failure and with acceptable oxygen saturation. Clinical and echocardiographic surveillance was planned, with repeat transthoracic echocardiography after one month.

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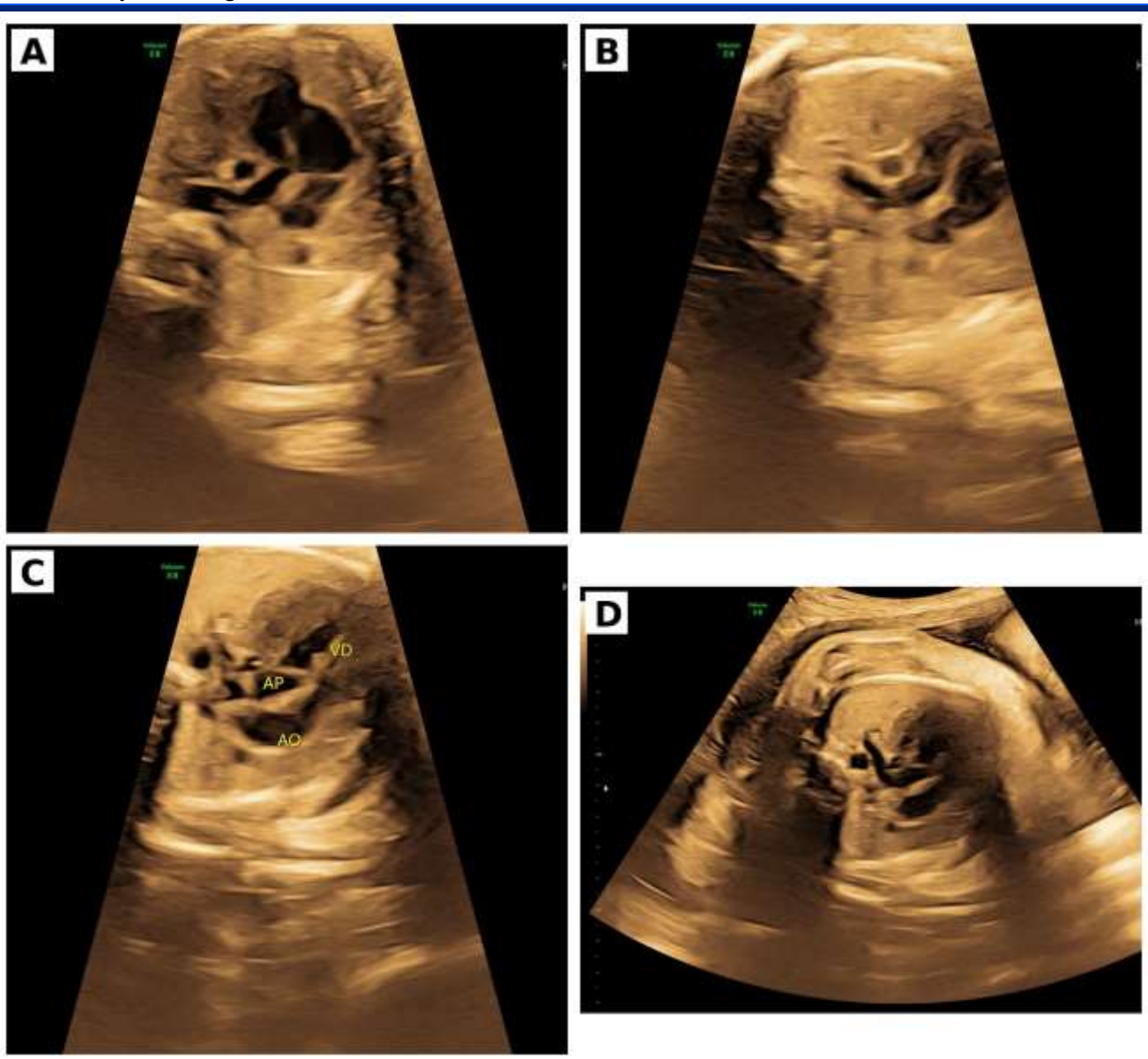

Fig. 1. De-identified antenatal fetal cardiac ultrasound at 38 weeks and 3 days. (A, B) Four-chamber and transverse cardiac views showing ventricular imbalance and suspected ventricular septal defect. (C) Outflow-tract view showing the right ventricle (VD), aorta (AO), and pulmonary artery (AP) with abnormal spatial relationship. (D) Additional axial cardiac view supporting the diagnosis of a complex conotruncal anomaly. AP: pulmonary artery; AO: aorta; VD: right ventricle.

DISCUSSION

DORV is defined by the origin of both great arteries predominantly from the morphological right ventricle [1,2]. Its anatomical spectrum is wide, and clinical presentation depends mainly on the relationship between the ventricular septal defect and the great arteries, the presence or absence of pulmonary stenosis, the degree of ventricular imbalance, and associated atrioventricular or extracardiac anomalies [3,4].

The diagnosis of DORV requires a systematic segmental analysis of the fetal heart. The four-chamber view may suggest associated abnormalities such as ventricular imbalance, atrioventricular septal defect, or valve malformation. However, the essential diagnostic step is assessment of the outflow tracts and precise identification of the origin of the great arteries [3]. Conventional two-dimensional fetal echocardiography can be sufficient when performed by experienced operators, but distinction from other conotruncal anomalies may remain difficult, particularly in late gestation or when fetal position is unfavorable [3,4].

In the present case, antenatal ultrasound detected a complex cardiac malformation with ventricular septal defect, malposed great arteries, abnormal ventriculoarterial connection, and pulmonary outflow tract narrowing. The

presence of two great vessels apparently arising from the right ventricle, with an anterior aorta and a posterior pulmonary artery, supported the diagnosis of DORV associated. The narrow subpulmonary pathway and smaller pulmonary artery suggested associated pulmonary stenosis. Postnatal echocardiography confirmed the prenatal suspicion and refined the anatomical diagnosis by demonstrating DORV, atrioventricular canal defect, common atrioventricular valve, right ventricular dominance, moderately hypoplastic left ventricle, and valvular and supravalvular pulmonary stenosis.

Van Praagh et al. proposed a segmental classification of DORV according to the cardiac segments involved [5]. Group I includes DORV with conotruncal anomalies only, mainly involving the great arteries. Group II includes DORV associated with malformations of the atrioventricular canal and the ventricles. Group III includes DORV associated with heterotaxy syndromes and situs abnormalities, involving the great arteries, ventricles, and atria [5,8]. The present case is consistent with a complex Group II pattern because DORV was associated with atrioventricular canal defect, ventricular imbalance, and abnormal great artery relationship.

The differential diagnosis includes transposition of the great arteries, tetralogy of Fallot, truncus arteriosus, and complex ventricular septal defect with overriding great artery [3,4]. In transposition of the great arteries, the aorta usually arises from the right ventricle and the pulmonary artery from the left ventricle, creating two parallel circulations. In DORV, both great arteries arise predominantly from the right ventricle. In tetralogy of Fallot, the aorta overrides a ventricular septal defect but usually does not fulfill the criterion of both great arteries originating from the right ventricle. Therefore, accurate analysis of ventriculoarterial connections is fundamental for diagnosis and postnatal planning.

The prognosis of DORV is highly variable and depends on anatomical subtype and associated lesions. Pulmonary stenosis may limit pulmonary overcirculation and explain the relative clinical stability of some neonates. However, severe pulmonary stenosis may contribute to cyanosis and, in more severe forms, ductus-dependent pulmonary blood flow. Adverse prognostic factors include ventricular hypoplasia, multiple ventricular septal defects, aortic arch obstruction, severe atrioventricular valve regurgitation, and extracardiac or chromosomal anomalies [3,4,6].

Genetic evaluation is an important component of the work-up of complex congenital heart disease. DORV has been associated with chromosomal abnormalities, including trisomy 13, trisomy 18, and chromosome 22 abnormalities [1,2,8]. A previously reported case described a complex DORV associated with a de novo pericentric inversion of chromosome 22 and deletion of the 22q12 region, suggesting possible implication of genes located in this region in early cardiac looping and DORV development [8]. In the present case, fetal karyotype or chromosomal microarray results were not available at the time of manuscript preparation. This represents a limitation, and genetic evaluation should be considered when feasible, especially in complex cardiac anatomy associated with severe fetal growth restriction.

This case also illustrates the importance of multidisciplinary perinatal management. Antenatal diagnosis allowed planned delivery in a tertiary center and immediate neonatal assessment. The pregnancy was also complicated by severe preeclampsia and severe fetal growth restriction, increasing maternal and fetal risk. Maternal stabilization and timely delivery were therefore essential, while neonatal cardiology assessment was anticipated immediately after birth. The favorable initial neonatal condition and absence of heart failure were probably related, at least in part, to pulmonary stenosis limiting pulmonary overcirculation.

The main strength of this report is the correlation between antenatal ultrasound findings and postnatal echocardiographic confirmation. The main limitations are the absence of fetal genetic testing, absence of long-term neonatal follow-up, and need for further evaluation to define the definitive surgical strategy. Longer follow-up will be necessary to assess clinical evolution, timing of intervention, and prognosis.

Conclusion

DORV is a rare and heterogeneous congenital heart disease requiring precise anatomical assessment. Prenatal ultrasound can suggest the diagnosis by identifying abnormal ventriculoarterial connections, ventricular septal defect, great artery malposition, and associated pulmonary outflow obstruction. In this case, antenatal imaging raised suspicion of complex DORV, later confirmed by postnatal transthoracic echocardiography as DORV associated with atrioventricular canal defect, ventricular imbalance, and pulmonary stenosis.

This case highlights the value of prenatal diagnosis in complex congenital heart disease, particularly for parental counseling, multidisciplinary delivery planning, and immediate neonatal management. Postnatal echocardiography remains essential to confirm the diagnosis, refine the anatomical classification, and guide follow-up and therapeutic planning.

Ethical Approval and Consent

This case report was prepared in accordance with ethical standards for publication of anonymized clinical observations. Written informed consent for publication of anonymized clinical information and ultrasound images was obtained from the patient. No identifying information is included in the manuscript or figures.

Conflict of Interest

The authors declare no conflict of interest.

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REFERENCES

[1] Golbabaei A, Majnoun MT, Aghaei Moghadam E, Rastgar A, Naemi M. Prenatal diagnosis of a rare case of double-outlet right ventricle with tricuspid atresia. J Tehran Univ Heart Ctr. 2022;17(1):38-40.

[2] Wu H, Cai A, Zhang Y, Sun W, Wang Y. Prenatal diagnosis of a fetal double-outlet right ventricle with tricuspid valve atresia, a straddling mitral valve, and a hypoplastic left ventricle. J Ultrasound Med. 2012;31:654-656.

[3] Lagopoulos ME, Manlhiot C, McCrindle BW, Jaeggi ET, Friedberg MK, Nield LE. Impact of prenatal diagnosis and anatomical subtype on outcome in double outlet right ventricle. Am Heart J. 2010;160:692-700.

[4] Obler D, Juraszek AL, Smoot LB, Natowicz MR. Double outlet right ventricle: aetiologies and associations. J Med Genet. 2008;45:481-497.

[5] Schallert EK, Danton GH, Kardon R, Young DA. Describing congenital heart disease by using three-part segmental notation. Radiographics. 2013;33:E33-E46.

[6] Brown JW, Ruzmetov M, Okada Y, Vijay P, Turrentine MW. Surgical results in patients with double outlet right ventricle: a 20-year experience. Ann Thorac Surg. 2001;72:1630-1635.

[7] Bhatla P, Tretter JT, Chikkabyrappa S, Chakravarti S, Mosca RS. Surgical planning for a complex double-outlet right ventricle using 3D printing. Echocardiography. 2017;34:802-804.

[8] L'Hermine-Coulomb A, Houyel L, Laroudie M, Aboura A, Audibert F, Dal Soglio D, et al. Ventricule droit à double issue et chromosome 22: une nouvelle délétion. Ann Pathol. 2004;24:1S148-1S149.