Original Research Paper

Volume - 16 | Issue - 09 | September - 2026 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Anatomy

DOUBLE OUTLET RIGHT VENTRICLE – A CLINICAL CASE WITH LITERARY REVIEW

Dr. Daisy Dwivedi* Assisstant Professor, Dept. of Anatomy, Pt. Jawaharlal Nehru govt. medical college and hospital, Chamba, Himachal Pradesh*Corresponding author

Dr. Bal Chander Professor, Dept. of Pathology, Dr. Rajendra Prasad Govt. medical college and hospital, Kangra at Tanda, Himachal Pradesh

ABSTRACT **Objective:** Congenital cardiovascular malformations contribute to the maximum number of developmental birth defects as reported by various studies. However, with advancement of technology we have been better equipped for timely diagnosis and hence, fairer outcomes frequently. We came across a rare case of Taussig Bing syndrome or double outlet right ventricle (DORV) while performing routine fetal autopsy. The article underscores the importance of fetal autopsies and reporting of such rare congenital anomalies. **Clinical Case:** A 19week fetus was received from obstetrics and gynaecology department preserved in formalin for autopsy. During autopsy we came across the rare congenital malformation where the two great vessels were arising from the same chamber of the heart. Further dissection lead to the diagnosis of a case of double outlet right ventricle (DORV). **Discussion:** The average life span of child suffering from uncomplicated DORV after surgical and supportive intervention, has been reported to be up to 15 years in few cases. Timely intervention can bring out the difference in outcome, affecting the life of parents and the surviving child. **Conclusion:** Awareness of such cases with appropriate investigations and counselling can reduce morbidity, mortality and patient suffering.

KEYWORDS : Double outlet right ventricle (DORV), Taussig Bing syndrome, fetal autopsy, embryology, teratology

INTRODUCTION

Developmental failures of outflow tracts of ventricles along with great vessels are termed as conotruncal anomalies in embryology. These are complex malformations. Double outlet right ventricle (DORV), tetralogy of Fallot, transposition of great arteries and double outlet left ventricle are some of the conotruncal anomalies. Definition of DORV has evolved over time. It was first described in 1957 **(1)**. Owing to morphological overlap with transposition of great arteries, it was considered as partial transposition of great arteries (TGA) with only the aorta transposed. Subsequently, it was decided that for the diagnosis, at least 50% of valve diameter of both great arteries must arise from morphological right ventricle. the criteria for diagnosis was put on firmer grounds by Goor et al. gave the definition of DORV as the cardiac condition wherein both great arteries arise from the morphologic right ventricle (RV). **(2)** Since it is a malformation occurring during embryogenesis, the anatomic spectrum is rather wide with different configuration of atria, atrioventricular connection, both left and right ventricular morphology and spatial relationship between both great arteries. As a consequence, DORV may hemodynamically mimic ventricular septal defect (VSD), tetralogy of Fallot (TOF) and transposition of the great arteries (TGA). **(3)** Tetralogy of Fallot need to be distinguished from DORV. The latter displays extreme dextroposition of the aorta, as there is loss of fibro-membranous continuity between the aortic and mitral valves and the aorta arises completely from the right ventricle without any override of the ventricular septal defect. Absence of pulmonary artery overriding ventricular septal defect in DORV also differentiates it from transposition of great arteries. Again, the absence of override is of particular importance in distinction of DORV from the Eisenmenger complex and the Taussig-Bing malformation (Van Praagh, 1968). **(4)** DORV also poses a challenge for clinicians due to wide anatomical spectrum. In the past few decades, advances in echocardiography and cardiac magnetic resonance imaging (MRI) have greatly enhanced diagnosis with ability to evaluate severity. **(5)**

Case report

We received a 19-week fetus from OBG department preserved in formalin after the consent of the parents. The fetus belonged to a 23-year primigravida with no significant medical history. There was no defect observed on gross examination. On performing the autopsy, the morphology of heart showed hypoplastic left ventricle with hypertrophy of left atrium with hypertrophy of left auricle as seen in the figure below. The sectioning of heart revealed aorta and pulmonary trunk arising from right ventricle. The aorta was situated on the right side of pulmonary trunk with d-malposition. The conal musculature under both the great vessels could not be differentiated due to muscular hypertrophy, neither a wide ventricular septal defect could be appreciated except for a slit like sub-pulmonary opening, which is usually the case in such anomalies. The right ventricular wall showed significant hypertrophy. The left ventricle was observed to have only a slit like thin cavity with dilatation of left atrium. The mitral and aortic valve were not in continuity. (Figure 1)

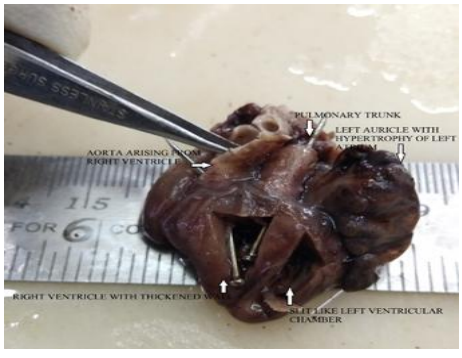

Figure 1: photograph showing both aorta and pulmonary trunk opening in right ventricle with enlarged right atrium encroaching on hypoplastic left ventricle. (white arrows)

DISCUSSION AND LITERARY REVIEW

DORV is rare and most children die at an early age owing to pulmonary hypertension. The prevalence has been reported to range from 0.7 to 2.7%. **(1)**

Definition of DORV has evolved over time as associated anomalies were noted and relative positions of great vessels were closely examined. Taussig and Bing described this entity as a "complete" transposition and left position of the aorta, with subpulmonic Ventricular Septal Defect also known as Taussig–Bing anomaly. **(6)** Later as more associated anomalies were noted, in 1957, DORV was classified by Witham into three groups: those with pulmonary stenosis (PS), those without PS, and those with an atrio-ventricular septal defect (AVSD) as well as other malformations **(7)** In the following years relative positions of great vessels were closely scrutinized and Maurice Lev pointed out the salience of position of the VSD, the great arteries, and their special relationship between each other. This is the predominant classification that is most frequently used to classify this congenital anomaly in the modern era. Crucial difference between TGA and DORV can be succinctly expressed in terms of concordant and discordant connection of great arteries with ventricles. In DORV PA is concordant since it arises out of RV and aorta is discordant since it arises out of RV in place of LV. TGA on the other hand shows discordant connections between arteries and ventricles. Some authors have professed the importance of relative postions of aorta and PA to each other. Prefixes "d" (right) or "l" (left) in the setting of DORV is used depending on aorta being positioned to the right or left of the pulmonary trunk is reffered to as d-malpositioned and l-malpositioned, respectively **(8)**. In the present case aorta was positioned right to pulmonary artery.

One of the largest studies involving of 27 cases of DORV was carried out by Cameron et al. All except three cases (11.1%) showed left ventricular hypoplasia. Slight, moderate and severe hypoplasia was seen in 6 (22.2%, 8 (29.6%) and 8 (29.6%) cases respectively and one case (3.7%) was found to be hypertrophic by the authors. 11 (40.7%) died within first month of life, 12 (44.4%) between one month and one year and 4 (14.8%) cases died after one year. Mitral valve was normal in 4 (14.8%), atresia was reported in 13 (48.1%), hypoplastic in 4 (14.8%). In rest 4 (14.8%) was either straddling or moot in cases 2 (7.4%) of common atrioventricular canal. **(4)** Present case also showed dilated left atrium and auricle, although we were not able to ascertain mitral atresia and the valve appeared to be normal compared to hearts of other equivalent gestational age fetuses.

Trisomy 13 and 18 have also been seen to be associated with DORV with hypoplasia. Trisomy 21 has also been shown in approximately 4% of the cases of DORV. We did not have the opportunity to harvest viable cells for karyotyping, however absence of brain, spinal, renal and facial abnormalities in the present case plausibly exclude such trisomies. **(2)**

There are occasional case reports of cases such as the present one surviving into adulthood. A 38year old man with total situs inversus, markedly hypoplastic left ventricle, moderate subpulmonary stenosis and DORV was reported. **(1)** Similarly there is a case of a 16-year-old female with DORV and two VSDs **(5)**

CONCLUSION

The case is being presented to detail the malformation and also to underscore the importance of fetal autopsies. Fetus had died in utero. Given the mortality rate and rather severe morbidity associated with such cases, parents as well as treating physicians are faced with a dilemma regarding continuation of pregnancy particularly when the diagnosis is made in third trimester. In a developing country like India with limited resources it may not be imprudent to terminate the pregnancy.

REFERENCES

1 Arenas-Fabbri V, Andrade-Cuellar EN, Guillot-Castillo SY, Monroy-Jiménez MA, Maldonado-Tenesaca AP, Aceves-Millan R, Solis-Gómez JC, Robledo-Nolasco R, de Jesús Encinos-Méndez D, Elizalde-Uribe IA. Double-Outlet Right Ventricle in an Adult With a Univentricular Heart and Total Situs Inversus. JACC Case Rep. 2025 Jul 9;30(18):103958. doi: 10.1016/j.jaccas.2025.103958. PMID: 40645705; PMCID: PMC12441456.

2 Bell-Cheddar Y, Devine WA, Diaz-Castrillon CE, Seese L, Castro-Medina M, Morales R, Follansbee CW, Alsaied T, Lin JI. Double outlet right ventricle. Front Pediatr. 2023 Sep 25;11:1244558. doi: 10.3389/fped.2023.1244558. PMID: 37818164; PMCID: PMC10560996

3 Goo HW. Double Outlet Right Ventricle: In-Depth Anatomic Review Using Three-Dimensional Cardiac CT Data. Korean J Radiol. 2021 Nov;22(11):1894-1908. doi: 10.3348/kjr.2021.0248. Epub 2021 Sep 13. PMID: 34564964; PMCID: PMC8546142

4 Cameron AH, Acerete F, Quero M, Castro MC. Double outlet right ventricle. Study of 27 cases. Br Heart J. 1976 Nov;38(11):1124-32. doi: 10.1136/hrt.38.11.1124. PMID: 1008954; PMCID: PMC483144

5 Khalil I, Abrar S, Hossain MI. Unraveling the complexities of double outlet right ventricle (DORV): A rare case of congenital heart disease with bidirectional shunting in a young girl. Radiol Case Rep. 2025 May 22;20(8):3930-3937. doi: 10.1016/j.radcr.2025.04.075. PMID: 40496077; PMCID: PMC12149962.

6 Taussig HB, Bing RJ. Complete transposition of the aorta and a levoposition of the pulmonary artery; clinical, physiological, and pathological findings. Am Heart J. (1949) 37:551–9. 10.1016/0002-8703(49)91133-3

7 Witham AC. Double outlet right ventricle; a partial transposition complex. Am Heart J. (1957) 53:928–39. 10.1016/0002-8703(57)90329-0.

8 Martins P, Castela E. Transposition of the great arteries. Orphanet J Rare Dis. (2008) 3(27). 10.1186/1750-1172-3-27