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Original Research Paper

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A RARE NEONATAL CARDIAC CASE OF DOUBLE OUTLET RIGHT VENTRICLE WITH ASD AND PAH.

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ABSTRACT **Objective:** Congenital Heart Diseases (CHD) are the most common structural birth abnormalities. A Neonate presenting in the first month of life without cyanosis, but with clinical features of congestive cardiac failure (CCF) and murmur on auscultation needs to be further investigated for lesions like a large VSD with PDA, unobstructed TAPVC, Double Outlet Right Ventricle (DORV) and Single Ventricle. **Background:** The foetal heart begins to form on 18th day of pregnancy, with formation of a heart tube that elongates and develops dilation and contractions ability as a ventricle and outflow tract. Both arterial trunks arise from right ventricle. Persistence of left ventriculo-infundibular fold (VIF) between aorta and mitral valve, which was supposed to be regressed is associated with DORV. By fourth and fifth week of foetal life atrial septum and ventricular septum develop. These structural developments are affected by idiopathic, genetic and environmental factors; resulting into CHD in-situ. But the presentations vary with lesion, age and clinical representation. The incidence being 6-8 per 1000 live births. It is most common direct cause of death in paediatric age group due to congenital heart anomalies, if not investigated and treated within time. **Discussion:** DORV is a complex CHD where aorta and pulmonary artery arise predominantly from morphological right ventricle along with presences of Atrial Septal Defect and Ventricular Septal defect (VSD) which requires a definitive surgical management. If DORV is present with cyanosis, there is pulmonary stenosis (PS) which resembles a Tetralogy of Fallot. In contrast, our case has pulmonary hypertension without cyanosis, but with features of CCF, as seen in DORV without PS. **Take Home Message:** With advancements in foetal and paediatric cardiology, early diagnosis and treatment of CHDs is now possible, and is of utmost importance for survival, prevention of complications, genetic counselling and improving the quality of life.

KEYWORDS : Double Outlet Right Ventricle, Ventricular Septal Defect, Atrial Septal Defect, Patent Ductus Arterosus.

INTRODUCTION:

Double outlet right ventricle is a rare cardiac condition with incidence of 1 in 3,000. [1] The International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD) has defined DORV as "the hearts with both arterial trunks supported predominantly by underlying morphologic RV" where both great arteries arise entirely or predominantly from the morphological RV. [2]

Case Presentation:

A 23-day old female born to primigravida mother with normal antenatal period. Neonate was breech in presentation, born by spontaneous vaginal delivery with birth weight of 1.320 kg and gestational age of 34-35 weeks by New Billard Scoring. Neonate is born by consanguineous marriage of 2nd degree with pedigree chart in fig (1). Neonate CIAB but had breathing difficulty, grunting, lethargic for which she had NICU admission for 21 days in view of RDS with LBW and later referred to Vedantaa Hospital for further management. The neonate presented to us with complaints of difficulty in feeding, suck-rest-suck cycle, sweating over forehead and without bluish discoloration of skin since birth with increased symptoms since past few days.

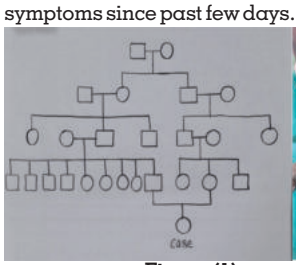

Figure (1)

Figure (2)

On general examination neonate had failure to thrive, tachycardia, tachypnea, maintaining saturation on 2L direct oxygen through nasal prongs, differential saturation in all four limbs was normal, Anterior fontanelle is at level 2 finger open while posterior fontanelle permits tip of finger, presences of pallor, periorbital edema, no cyanosis, CRT < 3 sec, peripheral pulsation are normal, good cry, tone and activity. Anthroprometrically neonate is (under minus) <-3 SD for weight for age, height for age and head circumference. All vitals were within normal limit.

The detailed systemic examination was done. In cardiovascular system examination the heart rate is 168 beats/min, apex impulse in 5th intercoastal space and left parasternal impulse in 2nd and 3rd Intercoastal Space, thrill in left parasternal region, pansystolic murmur grade 4//6 in left parasternal and infraclavicular region with radiation all over the chest and loud P2 with signs of CCF like Liver and spleen 3cm palpable below subcoastal margin, soft in consistency, round borders while, in respiratory system presence of respiratory rate of 84 cycles/min, intercoastal and subcoastal retractions and no basal rales. Rest other systems are normal.

On further investigating the child on chest x-ray cardiomegaly was noted figure (3) and 2D echography confirmed Double outlet right ventricle with overriding of Aorta, large subaortic 6.7mm inlet VSD with bidirectional shunt, normally related great arteries, normal biventricular function, normal pulmonary valve, no pulmonary stenosis and hyperkinetic PAH (RSVP 50mmHg + RA Pressure). 5.mm ASD shunting left to right. Figure (4,5,6,7 and 8)

Hence the diagnosis of Double outlet right ventricle with ASD

and PAH was established based on the in-depth history, examination and investigation findings.

Figure (3): Chest X-Ray

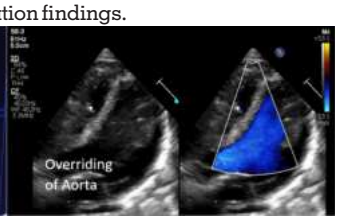

Figure (4): 2D Echography (overriding of Aorta)

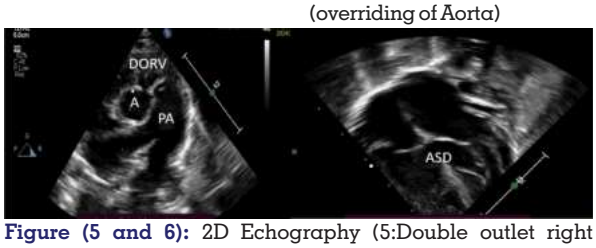

Figure (5 and 6): 2D Echography (5:Double outlet right ventricle is seen with aorta and pulmonary artery,6: atrial septal defect)

The child on admission was started on with medical management of Inj Lasix 0.5mg/kg/dose daily on alternate days and gradual introduction of feeds with vati spoon. 1-pint PCV was given as Hb was decreased but rest parameters were within normal range. After 3 days of Inj Lasix there was decrease in periorbital swelling and span of liver. Neonate was referred to cardiologist for opinion where 2D echography was done, syp furoped was advised till surgery and feed volume was advised not to be increased beyond 20ml. Further surgical management of VSD Closure was planned after 2 months along with attainment of 1.8 -2kilograms of weight by neonate.

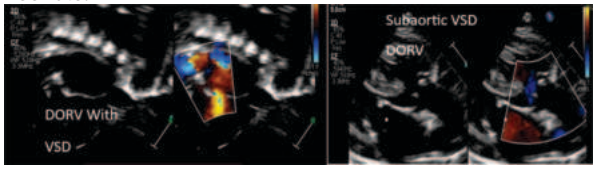

Figure (7 and 8): 2D Echography (7: DORV with VSD and 8: Sub-Aortic VSD with DORV)

DISCUSSION:

Due to genetic, environmental and various unknown causes affecting the fetal development result in the structural abnormalities occur in the heart or great intrathoracic vessels lead to congenital heart disease. It is the most common type of birth defect and cause of death in pediatric age group. Congenital heart disease is further divided into cyanotic also known as critical congenital heart disease (CCHD) and acyanotic congenital heart disease. Critical congenital heart disease has further three division: left sided obstructive lesions, right sided obstructive lesions and mixed lesion. Congenital heart disease is seen in 8-9 per 1000 live births. [3]

Normally in heart Aorta carrying oxygenated blood arise from the left ventricle whereas pulmonary artery carrying deoxygenated blood arise from right ventricle.[1] Double outlet right ventricle (DORV) is defined as the cardiac condition where both the aorta and pulmonary artery arise from the right ventricle, usually associated with ventricular septal defect or atrial septal defect.[2] In our case the neonate has double outlet right ventricle with both atrial and ventricular septal defects (VSD). In DORV if there is a subaortic presences of the VSD the left ventricle has an outlet via the septal defect into the aorta giving us the picture of overriding of the aorta and causing mixing of oxygenated and deoxygenated blood going towards the body. [1,6]

DORV belongs to a spectrum of conotruncal abnormalities from tetralogy of Fallot to complete transposition of great vessels. The clinical picture of DORV is same as of tetralogy of Fallot, if DORV is present with subaortic VSD with pulmonary stenosis. [4,5] The incidence of double outlet right ventricle is 0.03 to 0.14 to 1000 live births. [6]

In case pulmonary stenosis is present in DORV clinically there will be cyanosis and if no pulmonary stenosis, then neonate will have features of congestive cardiac failure. In our neonate despite the presences of pulmonary arterial hypertension there was no cyanosis and 2D echo report being the confirmatory to the diagnosis. [1]

For genetic counselling and prevention of further complication, early diagnosis and treatment is of utmost importance. Taking an in-depth history of complaints, birth, family and nutritional is noteworthy. Anthropromterical measurements of weight for age, height for age, head circumference and chest circumference help in keeping the track of growth in the neonate. In order to plan and implement the nursing care assessment of vital signs, oxygen saturation, skin colour (pink, cyanotic, mottled), mucous membrane (dry or cyanotic), peripheral pulse (rate, symmetry, quality), oedema, capillary refill, cold to touch, respiratory pattern, heart sound, feeding behaviour, intake and output, etc. Surgical procedures can be planned depending upon the age of presentation, severity of complaints, weight, type of VSD and aberrant coronary artery distribution. [6]

Conflict Of Interest: No conflict of Interest.

REFERENCES:

1. Chowdhury AR, Punj J, Kayina CA, Singh A, Jasiel JG. Anesthetic Management of a Parturient with Double-Outlet Right Ventricle Obstruction and Pulmonary Stenosis Scheduled for Caesarean Delivery: Case Report and Review of Literature. AANA journal. 2021 Dec 1;89(6):523-8.
2. Bell-Cheddar Y, Devine WA, Diaz-Castrillon CE, Seese L, Castro-Medina M, Morales R, Follansbee CW, Alsaied T, Lin JH. Double outlet right ventricle. Frontiers in Paediatrics. 2023 Sep 25; 11:1244558.
3. Galvis MM, Bhakta RT, Tarmahomed A, Mendez MD. Cyanotic heart disease. InStatPearls [Internet] 2023 Jan 31. StatPearls Publishing.
4. Weir EK, Joffe HS, Barnard CN, Beck W. Double outlet right ventricle: clinical and anatomical spectrum. Thorax. 1978 Jun 1;33(3):283-9.
5. Sondheimer HM, Freedom RM, Olley PM. Double outlet right ventricle: clinical spectrum and prognosis. The American Journal of Cardiology. 1977 Jan 1;39(5):709-14.
6. Waghaye M, Sakharkar S, Pathade A. CASE REPORT ON TETRALOGY OF FALLOT (TOF) WITH DOUBLE OUTLET RIGHT VENTRICLE (DORV). Journal of Pharmaceutical Negative Results. 2022 Oct 1;13(3):927-30.