A Case Of Double Outlet Right Ventricle With Extremely Rare Anatomical Features

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Abstract: Double outlet right ventricle (DORV) is a rare cardiac malformation especially in adulthood. We report a 27-year-old man with DORV along extremely rare anatomical features. The diagnosis of DORV in this case was made with echocardiography, also the surgical treatment for the case have been discussed. [Patel M Natl J Integr Res Med, 2020; 11(3):88-90]

Key Words: double, outlet, right, ventricle, ventricular, septal, defect

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Introduction: Double outlet right ventricle (DORV) is found in a group of complex heart lesions, which are unified by the characteristic that both great arteries arise predominantly from the right ventricle. The physiology of the DORV after birth is determined mainly by the location of the ventricular septal defect (VSD) in relation to the great arteries as well as the presence or absence of outflow tract obstruction[1]

Case Report: A 27-year-old male patient presented to the emergency room of a tertiary care clinic in Ahmedabad, Gujarat India with chief complaint of dyspnea on rest. The general physical examination was performed. The patient was well alert and oriented to time, place and person. His vitals were taken which recorded a pulse rate of 96 beats per minute, in the right radial artery. His pulses were symmetrical and had normal amplitude. On physical examination, the patient had tachypnea and appeared cyanotic, with clubbing of fingers and toes along with facial edema. Patient gave a past history of multiple hospital visits for the complaints of growth retardation and breathlessness, but no diagnosis was made.

Further, cardiovascular examination was performed, during auscultation the following results were obtained— S2 was loud and single, mid systolic murmur (ejection) along the left sternal border was present.

His laboratory investigation reports are as follows- hemoglobin (Hb: 19.4gm/dl) indicating polycythemia, increased levels of hepatic enzymes due to congestive liver and marked hypoxemia with Partial pressure of oxygen- (pO2)- 50 mmHg. Further, Chest X ray- thorax- was performed. The chest X ray revealed an enlarged cardiac silhouette due to enlarged right cavities without increased pulmonary vascular markings. His Electrocardiogram showed right ventricular hypertrophy.

Echocardiography was found to be the best diagnostic method for the patient. The echocardiography revealed the following findings: situs solitus, blood from Superior Vena Cava (SVC) and Inferior Vena Cava (IVC) draining into right atrium to right ventricle, from the right ventricle blood going to aorta and pulmonary arteries, from the pulmonary veins oxygenated blood draining into the left atrium to left ventricle and from the left ventricle it was going to the right ventricle through sub aortic Ventricular Septal Defect (VSD). Right atrium and right ventricle were found dilated with left ventricular ejection fraction (38%) with normal left ventricle size. Infundibular stenosis was found at the pulmonary valve. Normal relationship was found in the great arteries.

Discussion: In cases of double outlet right ventricle, the hearts can present either a subaortic VSD or a sub pulmonary VSD, the VSD in both forms is found between the limbs of the TSM [2,3]. It is found that the specific attachment of the infundibular septum to the anterior limb of the TSM that makes the VSD subaortic. When the infundibular septum is attached to the posterior limb of the TSM, the VSD is then sub pulmonary.
In the patients of the hearts with a subaortic VSD, the distance between the aortic valve and the VSD is determined in part by the length and development of the right-sided VIF. The spatial relationships between the great arteries tend to be normal when the VSD is subaortic. side by-side great arteries with the aorta to the right of the main pulmonary trunk is the most common spatial relationship when the VSD is sub pulmonary (Taussig-Bing form) [4,5].

There are a few definite surgeries for the cases of double outlet right ventricle. These are:

1. Subaortic Or Doubly Committed VSD: An intraventricular tunnel between the VSD and the subaortic outflow tract is created by means of a Dacron patch. This procedure is performed early in life, preferably during the neonatal period or at least in early infancy, without preliminary PA banding.Sometimes the RVOT may have to be augmented with an outflow patch if the VSD–AO tunnel obstructs the RV outflow tract. The mortality rate is less than5% for simple subaortic VSD; it is slightly higher for doubly committed VSD.

2. Fallot Type: There are three surgical options. Surgical repair is generally advised by 6 months of age, preferably during the neonatal period. However,if the patient's condition is poor or if there are major associated noncardiac anomalies, an initial shunt operation is an option.

a. Tunnel VSD closure + Rastelli operation: An intraventricular tunnel between the VSD and the aorta is established, and a Rastelli operation is performed to relieve PS using either a pulmonary or aortic homograft conduit.

b. REV procedure: So-called reparation a l'etage ventriculare (REV), which is similar to ASO, may be performed. In the REV procedure, the proximal ascending aorta and main PA are transected, and the proximal stump of the PA is over sewn. The pulmonary arteries are trans located anterior to the aorta (Lecompte maneuver), and the ascending aorta is reconnected. The distal PA is anastomosed directly to the upper margin of the infundibular incision. Autologous pericardium forms the anterior portion of the pathway. The hospital mortality rate is 18%.

c. Nikaidoh procedure: This combines the principle of the Ross procedure and the Konno operation. The aortic root including the aortic valve is detached in the same manner as done to the pulmonary root in the Ross procedure. The PA is divided, and the pulmonary valve is excised. The pulmonary root is divided, and the conal septum above the VSD is excised, which creates a large opening to the LV cavity. The aortic root is trans located posteriorly and sutured to the open orifice of the pulmonary annulus. A pericardial patch is used to connect the lower margin of the VSD and the anterior circumference of the harvested aortic root, completing the LV to AO connection. A pericardial gusset completes the connection of the RV and the distal end of the MPA.

3. Taussig-Bing Anomaly (Subpulmonary VSD): There are four possible surgical approaches. These operations should be carried out by 3 to 4 monthsof age or sooner because of the rapid development of pulmonary vascular obstructive disease in this subtype.

a. The procedure of choice is the creation of an intraventricular tunnel between the VSD and the PA (resulting in TGA), which is then corrected by the ASO. The mortality rate is between 5% and 15%.

b. Creation of an intraventricular tunnel between the VSD and the PA is followed by the Senning operation. This is a less desirable approach because of a high mortality rate (>40%) and a higher late complicationrate associated with the Senning procedure.

c. An intraventricular tunnel between the VSD and the aorta is desirable but often technically impossible. The surgical mortality rate is about 15%.

d. Creation of a VSD-to-PA tunnel followed by the Damus-Kaye-Stansel operation and RV–to-PA conduit is another possibility.

4. Non Committed VSD: When possible, an intra ventricular tunnel procedure between the AV canal-type VSD and the aorta is performed, but the mortality rate is high (30%–40%). PA banding is usually needed in infancy to control CHF, and the surgery may be delayed until 2 to 3 years of age.

Conclusion: Double outlet right ventricle can rarely in the hospitals and can be quickly diagnosed with echocardiography and treated promptly by surgical intervention.

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