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Double Outlet Right Ventricle: Conventional and Fuwai Classification and Guidance for Surgical Correction

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ABSTRACT

The Double Outlet Right Ventricle (DORV) is a complex congenital heart disease and its surgical correction is a difficult task for the surgeon. Preoperative evaluation and categorization of DORV sub types by modified fuwai criteria can give prerogative guidance to the surgeon for the reconstructive surgery of DORV. In this retrospective study we compared modified fuwai classification of DORV sub types and surgical correction of different sub types. Male gender was predominant 61%. Majority of the patient was in fuwai class IB (56.25%) and fuwai class IA (16.6%). In fuwai IB subtypes of DORV required tunnel creation, Right Ventricular Outflow Tract Obstruction release (RVOTO) and pulmonary valve repair (40%) and in class I A required left ventricle to aortic tunnel creation (15%). In IIA required VSD enlargement and tunnel creation (6%) and in type IIB required interventricular tunnel repair, pulmonary valve creation and palliative surgery (11%). Our conclusion is preoperative Fuwai classication can guide surgeon about the surgical approach in DORV.

Keywords: DORV; Fuwai classification; DORV surgery; DORV classification

INTRODUCTION

DORV is a ventricular arterial anomalies, is a complex congenital heart disease. Where more than 50% of both aorta and pulmonary artery arise from the morphological right ventricle. There is heterogeneous cardiac malformations are found in DORV. But common morphologic feature is the origin of both great arteries from the morphologically right ventricle. There is variable amount of canal tissue present below the semilunar valve. The reported incidence of DORV 0.6 cases per 10000 live births and there is no sex variation.

during echocardiographic evaluation of DORV the 4 important findings should be specified (i) Anatomical location of Ventricular Septal Defect (VSD) in relation to Great Arteries (GA), (ii) Size of the VSD, (iii) Relationship of the GA and (iv) Presence or absence of outflow obstruction (RVOTO). These echo cardio graphic findings classify the DORV in conventional classification as normal GA relation and abnormal GA relation (Side by side, D-malposed, L-malposed) with VSD location (sub aortic, sub pulmonary, doubly committed and remote) and another classification is modified fuwai classification, proposed. Where DORV was classified on

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basis of three parameters like GA relation, location of the VSD, and presence or absence of RVOTO. They classified DORV in 8 8 sub types (Table 1) [1-3].

Table 1. Modified fuwai classification with DORV.

DORV subtypes	Relative position of great arteries	Relation between GA and VSD	RVOTO
IA	Normal	Committed	Absent
IB	Normal	Committed	Present
IIA	Normal	Non committed	Absent
IIB	Normal	Non committed	Present
IIIA	Abnormal	Committed	Absent
IIIB	Abnormal	Committed	Present
IVA	Abnormal	Non committed	Absent
IVB	Abnormal	Non committed	Present

The modified fuwai classification gives a complete guidance to the surgeon about surgical procedure rather than conventional classification. In this study our objective was to categorize the patient with DORV according to conventional and modified Fuwai classification system and to see the surgical approach done by surgeon according to modified fuwai subtypes of DORV.

MATERIALS AND METHODS

This was a retrospective study conducted in the national heart foundation hospital and research institute, Darus salaam, Mirpur, Dhaka, Bangladesh. During the period of 2017 to 2021, all the patients who were underwent surgery for DORV were included and a total of 107 patients were operated for DORV. Preoperative echocardiographic, cardiac CT and cardiac catheter data and intra operative findings were used to classify the patient. Patient were classified on the basis three criteria VSD location (Committed or non-committed), GA relation (normal or abnormal relation), RVOTO (obstruction present or absent). Patient was sub categorized fuwai class-I am if committed VSD, normal GA relation and no RVOTO and IB if committed VSD plus Normal GA relation with RVOTO. IIA: Noncommited VSD plus normal GA and no RVOTO; IIB noncommittal VSD nor GA with RVOTO, IIIA: Commited VSD plus abnormal GA no RVOTO. IIIB: Committed VSD abnormal GA plus obstructed RVOT; IVA: Non committed VSD abnormal GA and no RVOTO and IVB non-committed VSD plus abnormal GA relation plus presence of RVOTO. The data of surgical procedure were also collected in predefined format. Finally types of surgery were comparing with modified Fuwai classification.

Data Analysis

Data analysis was done by using statistical software, SPSS package for window 10, vision 25, and data was checked for normal distribution. Skewed data was presented in mean and median with minimum and maximum range. Qualities and categorical date are presented in frequency and proportion. DORV subtypes and surgical procedures were compared by cross tabulation [4,5].

RESULTS

Among the 107 patients of surgically corrected DORV, male were predominant (67%) and mean age at diagnosis was 5.13 years and median age was 2.92 years. Age at prostration was not normally distributed and data was right skewed (Table 2).

Table 2: Demographic variable of the study population.

Variable	N (%)
Male	66(60.6)
Female	38(39.4)
Age (years)	
Mean	5.13
Median	2.95

Minimum	0.16
Maximum	30
Height (cm)	
Mean	94.46
Min-Max	51-164
Median	85.5
Body surface area	0.60 ± 0.32

Among the DORV subtypes TOF variety was the main subtypes (46.8%) and VSD type 40.8%, single ventricle 7.3% and TGA type was 2.8% (Table 3).

Table 3: DORV types in study population.

DORV Type	N (%)
TOF	51 (46.8)
TGA	5 (2.8)
VSD	44 (40.8)
Single ventricle	8 (7.3)
Situs inversus	1 (0.9)

In the view of VSD location 68% was perimembraneous type, sub aortic was 11%, sub pulmonic 3% and inlet type VSD was 9% (Table 4).

Table 4: VSD location.

VSD location	N (%)
Doubly committed	4 (1.80)
Inlet	10 (9.2)
Perimembranous (PM)	74 (67.9)
Pulmonary atresia	5 (4.6)
Sub aortic	12 (11)
Sub pulmonic	3 (2.80)
TOF	1 (0.9)

Conventional classi ication of DORV with relation of Great Arteries (GA) position and VSD location, the main category was normal GA and perimembranous VSD in 66 patients (73.33%), normal GA with sub aortic VSD was 11 (12.2%), D-malposed GA and PM VSD was in 5 patients and L-malposed with inlet type VSD was found in 1 patient (Table 5).

Table 5: Conventional classi ication of DORV (relation of GA and VSD location).

	VSD location					
GA relation (%)	PM	Sub-Aortic	Sub-Pulmonary	Inlet	Pulmonary atresia	Doubly committed
Normal	66 (73.33)	11 (12.2)	1 (1.1)	6 (6.7)	5 (5.6)	1 (1.1)

Side by side	4 (50)	11 (12.5)	1 (12.5)	1 (12.5)	---	1 (12.5)
D-Malposed	5 (62.5)	--	1 (12.5)	1 (12.5)	1 (12.5)	---
L-Malposed	-----	----	----	1 (100)	--	--

PM: Perimembranous.

Modified Fuwai classification by Pang majority of our patient was in fuwai class IB, 51 patients (46.8 %) and 14 (13%) patient was in IA and 8 (7.47%) patients were in IIB (**Table 6**). VSD closure and commissuretomy was the main surgical approach in type IB and in IA and tunnel formation plus VSD closure and commissuretomy was done equally in both IA and IB. VSD closure and pulmonary valve creation was required in 17 patients. Fontana procedure was done IIIB patient. BD glann was done 10 patients was in class IVB (**Figure 1**).

Table 6: Fuwai class and surgical classification.

Fuwai class	Bi ventricular repair	Uni ventricular repair	Staged surgery
IA	LV to Ao tunnel (n=16)	__	__
IB	Tunnel+RVOTO release +PV cre (n=43)	__	1 MBT (n=1)
IIA	VSD enlargement +tunnel (n=7)	__	__
IIB	IVR+PV cre+palliative (n=12)	1	MBT (n=2)
IIIA	ASO+IVR(n=6)	__	
IIIB	PRT (n=1)	__	MBT (n=4)
IVA	IVR+ASO (n=1)	BD Glann (n=2)	__
IVB	__	Multiple patch, resection of band, re attachment of tricuspid valve chordae tendinae (n=12)	__

Ao-aorta PV cre: Pulmonary Valve creation; IVR: Interventricular Tunnel Repair; ASO: Arterial Switch; PRT: Pulmonary Root Translocation; MBT: Modified Blalock Tausig.

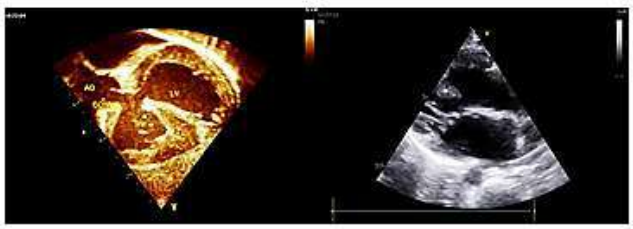

Figure 1: Parasternal long axis view aorto mitral discontinuity 90% overriding large VSD.

DISCUSSION

Our study findings suggest that modified fuwai classification can guide surgeon about preoperative decision about the surgical reconstruction and every DORV subtypes required specific surgical correction. This modified Fuwai classification was not used in our institute before this study. Our entire preoperative echo finding has the 100% accuracy with per operative finding this may be due to several time verification by different echo cardiographer. We did other imaging finding like cardiac CT angiogram and cardiac catheterization when necessary. Our study finding has similar statement with other study. We differentiate TOF type DORV (Type-IB) from TOF if over ridding is >50%. In type I-A DORV who had aortic mitral separation or long fibrous continuity and surgical approach was intra ventricular tunnel formation to Aorta and modification of surgical procedure was applied if any associated anomalies like restrictive VSD was present, and in I-B operative procedure was tunnel creation, RVOTO release and pulmonary valve creation. Type II-A was repaired with making long tunnel LV to aorta and VSD enlargement and tunnel wide was decided per operatively. Sometimes re implantation of tricuspid papillary muscle was required in type II-A. Pulmonary artery banding was done in both II-A and II-B, with RVOTO relief. Type III and type IV biventricular repair and re position of GA was done. In type III-B double root translocation was done as there was sub aortic coin and modification procedure done when there is ill defined coin with tunnel creation along with other corrective procedure if required like rastelli or right ventricular pulmonary artery conduit. In type IV-A like TGA had remote

VSD with both GA arises from the RV, biventricular repair to connect LV with PA and ASO was done. IV-B required multiple patches, resection of multiple band and reattachment of tricuspid valve's chordae endian when there was well sub aortic canal tissue; a double root translocation with tunneling of VSD to PA was done [6-8].

CONCLUSION

Majority DORV subtypes in convention classification normal GA and per membranous VSD and modified Fuwai classification type IB and surgical approach of DORV correction was VSD closure and commissuretomy in Fuwai type IA and IB. Pre surgical classification can guide surgeon about the surgical approach.

LIMITATION

Small sample and single center data. Preoperative data and Short term and long term outcome not shown in this study.

CONFLICT OF INTEREST

None

ETHICAL APPROVAL

It was approved by ethical review board of national heart foundation hospital and research institute.

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