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Original Article

Surgical Experiences and Results of the Management of Double Chambered Right Ventricle

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ABSTRACT

Objective: To identify anatomical features, association and surgical results of patient with double chambered right ventricle.
Study Design: Prospective longitudinal study.
Place and Duration of Study: The Children Hospital and Institute of Child Health, Lahore Pakistan, from Jan 2013 to Dec 2018.
Methodology: All patients presenting with mid cavity right ventricular outflow tract obstruction were included. All patients' demographic data, clinical profile, diagnostic reports, associated anomalies, and surgical data were recorded.
Results: Fifty-two patients with mid-cavity right ventricular outflow tract obstruction of various ages (from 6 months to 31 years) were included. Ten patients (19%) presented in infancy. Forty-six patients (89%) had an associated ventricular septal defect, ten (15%) had aortic valve right coronary cusp prolapse with varying degrees of aortic regurgitation. All patients had a right ventricular mid-cavity and infundibular muscle resection via the tricuspid valve. There was one hospital death due to an intraoperative global neurologic catastrophe. The median follow-up after surgery was 37.5 months. There was no late death.
Conclusion: Doubly committed Ventricular septal defect with aortic valve right coronary cusp prolapse with varying degree of aortic regurgitation and absence of subaortic stenosis is a new finding in our study. Early and Medium-term surgical results of repair are excellent.

Keywords: Aortic Insufficiency, Double Chambered Right Ventricle, Mid Cavity Right Ventricular Outflow Tract Obstruction, Right Coronary Cusp Prolapse

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INTRODUCTION

Congenital heart diseases (CHDs) are commonest of congenital defect having incidence of about 1-2 % and lead to a significant increase in burden of congenital diseases in children owing to high morbidity and mortality particularly in resource limited countries where surgical facilities are scarce and pose as major risk factors for adult cardiovascular problems.[1] It is estimated that about 400 genes are associated with CHD pathogenesis.[2] An anomalous muscle band divide the right ventricular cavity into two cavities, a high pressure lower cavity and low pressure upper cavity.[3] Majority around 90% of Double Chambered Right Ventricle (DCRV) patients have another associated congenital heart lesion. The patients are usually not cyanosed.[4,5] Right Ventricular cavity is essentially divided into two chambers by anomalous muscle bundles creating an intra-ventricular obstruction named as mid cavity right ventricular outflow tract obstruction (RVOTO). The diagnosis of most double chambered right ventricle is carried out in pediatric and adolescent patients, but rarely adult patients are also seen with this condition. DCRV patient will have variable symptom based on degree of obstruction such as exercise intolerance, easy fatigability, palpitation, breathlessness and cyanosis depending on severity of mid cavity obstruction.[5,6] The reported associated congenital cardiac anomaly in DCRV are Ventricular septal defect (90%) pulmonary valve stenosis (~40%), atrial septal defect (~17%), and double-outlet RV (~8%).[7] DCRV can also be an associated anomaly of various syndrome like Williams's syndrome.[8] Transthoracic Echocardio-graphy (TTE) is a first line diagnostic modality for the diagnosis of DCRV.[9] The need for invasive testing like cardiac catheterization is rarely required owing to high sensitivity and specificity of noninvasive diagnostic tool.[10] There is no defined parameter and timing of surgical intervention. We evaluated the anatomical features, associations and surgical results of patients undergoing surgery for this condition.

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METHODOLOGY

The prospective longitudinal study was conducted at the Department of Pediatric Cardiology and Cardiac Surgery, The Children Hospital and Institute of Child Health, Lahore Pakistan, from January 2013 to December 2018. Hospital IRB approval (2020-159-CH&ICH) was obtained. Non-probability consecutive sampling was employed.

Inclusion Criteria: Patients of either gender and age less than 18 years, who presented for elective surgery with transthoracic echocardiography-based diagnosis of Double chambered right ventricle showing mid-cavity Right ventricular outflow tract obstruction (RVOTO) due to obstructing bundles with a relatively normal infundibulum and pulmonary valve annulus with little anterior conal septal malalignment or proven on per operative finding of DCRV were included. Seven (13%) patients with the preoperative diagnosis of tetralogy of Fallot (TOF), so called "pink TOF" were regrouped as DCRV based on intra-operative findings were also included in study.

Exclusion Criteria: Patient with diagnosis of Tetralogy of Fallot, Isolated pulmonary valvular stenosis, and supra valvar pulmonary stenosis were excluded.

Preoperative evaluation with transthoracic echocardiography (TTE) was standard for all patients. All patients had an intra-operative epicardial echocardiography done to confirm adequate repair. A post resection intra-operative RVOT gradient of less than 25-30 mmHg was deemed to be acceptable. All patient's demographic data, clinical presentation, preoperative echo finding of DCRV, degree of RVOT obstruction, associated anomalies like size and location of Ventricular septal defect, abnormality of right coronary cusp prolapse, severity of aortic insufficiency, intraoperative finding like level and degree of mid cavity RVOT obstruction, location and nature of VSD, presence of RCCP, aortic insufficiency, surgical technique, immediate surgical outcome and follow up data were recorded on pre designed proforma. The information was entered into a structured database.

The results were evaluated using Statistical Package for social sciences (SPSS) Version 20. Gender, symptomatology, associated problem, surgical approach and procedure performed were presented as frequency and percentages. The chi square test and t-test were used for inferential statistics and p value of ≤ 0.05 was considered significant.

RESULTS

In our study sample, 52 patients aged 6 months to 31 years with a mean age of 60±58.9 months, underwent repair for Double chambered right ventricle. There were thirty-four (65%) male and 18(35%) female patients, and 10 patients presented in infancy (19%). Weight range was from 5.7kg to 82 kg, with mean weight being 15+11.47 kg. Forty-six patients (89%) had an associated VSD, ten patients (19%) had an aortic valve right coronary cusp prolapse with varying severity of aortic insufficiency (Figure-1). The median follows up range after surgery is 63.5 months with an interquartile range of 37.5 months. Commonest presenting complaints were Dyspnea (30, 57%), failure to thrive (11, 21%) and symptoms of heart failure (11, 21%). Patient having mid cavity Right ventricular outflow tract gradient exceeding 40mmHg, or right coronary cusp prolapse with or without aortic insufficiency and symptomatic patient were considered candidate for surgical repair. The median pre surgery right ventricular outflow tract gradient was 70mmHg (range 30 to 160 mm Hg) preo-peratively.

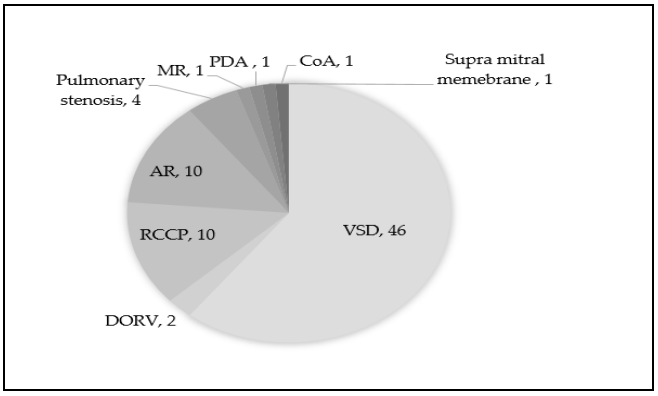

Figure-1 Associated condition with Double Chambered Right Ventricle (n=52)

Complete surgical repair was done through midline sternal incision in all cases using mild to moderate hypothermic cardiopulmonary bypass and cold ante grade del Nido blood cardioplegia for myocardial protection. The obstructing muscle bundles in the mid cavity Right Ventricular Outflow Tract were directly visualized intra-operatively. Operative approaches used are listed in Table-I.

Table-I: Operative Approaches in Double Chamber Right Ventricular Obstruction Surgery (n=52)

Operative Approach	n(%)
Trans-atrial	41(79%)
Trans-pulmonary-only	0(0%)
Trans-atrial/Trans-pulmonary	11(21%)
Trans-ventricular- only	0(0%)

Associated procedures performed included VSD closure (90%), aortic valve repair (11%), pulmonary valvotomy (8%) and 2% were PDA ligation, coarctation of aorta repair, mitral valve repair and supramitral membrane resection (Figure-2).

Age, gender, hospital stay, operative approach, symptomatology, isolated DCRV and those with other

associated congenital lesion had no impact on survival except for pulmonary stenosis which yielded a significant *p* value of less than <0.05. (Table II and III).

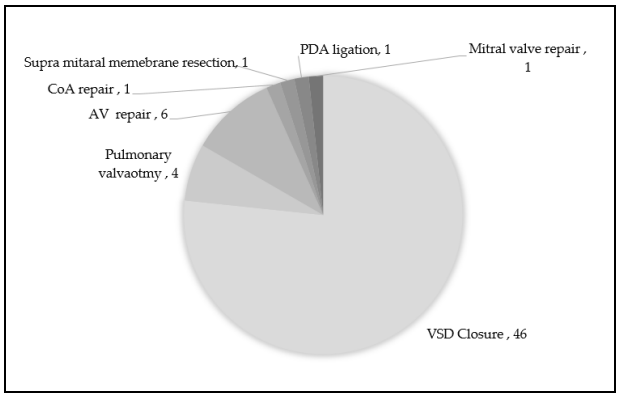

Figure-2: Surgical Procedures associated with Double Chambered Right Ventricle (n=52)

Table-II: Outcome for Age, Gender, Symptoms, Hospital Stay and Operative Approach (n=52)

Parameters		Outcome			*p*-value
		Death n (%)	Survived n (%)		
Age Group	Infants (Less than 1 year)	0(0.0%)	10(100.0%)		0.183
	Toddler (1-3 years)	1(8.3%)	11(91.7%)		
	Children and adult (More than 3 years)	0(0.0%)	30(100.0%)		
Gender	Male	1(2.9%)	33(97.1%)		0.463
	Female	0(0%)	18(100%)		
Hospital Stay	Equal to or less than 5 days	1(3%)	32(97%)		0.444
	More than 5 days	0(0%)	19(100%)		
Symptomatology	Dyspnea	0(0%)	30(100%)		0.150
	Failure to thrive	1(9.1%)	10(90.9%)		
	Heart failure	0(0%)	11(00%)		
Operative approach	Transatrial	1(12.5%)	41(87.5%)		0.302
	Transatrial + Transpulmonary	0(0%)	11(100%)		

Re-exploration for bleeding was required in 1(2%) patient, no patient required permanent pacemaker implantation or reoperation for residual VSD. Six (12%) patients needed an aortic valve repair for aortic insufficiency graded as being moderate to severe. No patient needed an aortic valve replacement. The residual Right ventricular outflow tract obstruction (RVOTO) required reoperation in 1(2%) patient in same admission. There were no late deaths.

There was a reduction of median RV gradient from 70mmHg (range 30-160mmHg) before surgery to median gradient of 12 mmHg (range 5-20) after surgical repair. The follow up transthoracic echocardiography showed significant reduction in RV muscle mass. Currently no patient who had aortic valve repair had progression of aortic valve insufficiency. 47 (90%) patients reported post-operative follow up.

Table-III: Outcome for Associated Lesions (n=52)

Associated Lesions	Outcome			*p*-value
	Death n(%)	Survived n(%)	Total n(%)	
DCSA VSD+RCCP +Mild AR	0	1(100%)	1(100%)	0.888
DCSA VSD + RCCP + Moderate AR	0	2(100%)	2(100%)	0.840
DCSA VSD + RCCP + Severe AR	0	2(100%)	2(100%)	0.840
DCSA VSD + RCCP + Severe AR + Supramitral membrane	0	1(100%)	1(100%)	0.888
DORV + PMVSD	0	2(100%)	2(100%	0.840
No associated lesion	0	4(100%)	4(100%)	0.771
PDA	0	1(100%)	1(100%)	0.888
PMVSD	0	29(100%	29(100%)	0.275
PMVSD + Coarctation of aorta	0	1(100%)	1(100%)	0.888
PMVSD + MR	0	1(100%)	1(100%)	0.888
PMVSD + valvar pulmonary stenosis	1(25%)	3(75%)	4(100%)	<0.001
PMVSD + RCCP + Mild AR	0	4(100%)	4(100%)	0.771
Valvar Pulmonary stenosis	0	1(100%)	1(100%)	0.888

DISSCUSION

Double-chambered right ventricle (DCRV) is a rare congenital heart anomaly in which abnormal muscle bundle divides right ventricular cavity into two cavities. The double chambered right ventricle incidence is 0.5% to 1 % of all congenital heart diseases, and occurs due to presence of hypertrophic anomalous muscle bundles.[6] It rarely occurs in isolation and after surgical repair of Ventricular septal defect and tetralogy of Fallot with a reported incidence of 10% and 6% post VSD closure and post tetralogy repair as reported by various studies.[11-14] All patients in our study were pediatric patient except one patient who is adult. All patient in our study were symptomatic which is also the result of previous studies which reported dyspnea or shortness of breath as commonest symptoms in about 36% cases followed by heart failure, fatigue and cyanosis each about 8%. In our study dyspnea and shortness of breath is also commonest symptoms in older patient (58%) while patient who present in infancy with mild mid cavity

obstruction and large left to right shunt along with patient with RCCP with moderate to severe aortic regurgitation have sign of heart failure (21%) and patient having severe mid-cavity obstruction with or without ventricular septal defect were failure to thrive in 1st two year of life (21%). None of our patients was cyanotic. Said SM *et al*. also reported the 15% incidence of heart failure in pediatric patients in his study. In our study one patient (2%) develop double chambered right ventricle obstruction after surgical correction of ventricular septal defect in infancy and five patients have isolated double chambered right ventricular obstruction and no patient have double chambered right ventricular obstruction post tetralogy repair. Moran reported an incidence of development of double chamber right ventricle in about six (6%) of patients who underwent surgical repair for tetralogy of Fallot before two years of age. In our study we have seen seven patients with diagnosis of tetralogy of Fallot who were missed preoperatively to have mid cavity right ventricular obstruction. These seven-patient found to have concomitant double chambered right ventricle obstruction on intraoperative echocardiogram requiring surgical resection of mid cavity obstructing muscle bundle and hence tetralogy of Fallot become the most common concomitant diagnosis (13%) with double chambered Right ventricle in our study.

There are reports of the diagnosis of DCRV being missed due to echo imaging limitations in adults.[10] Most adult patients in older studies underwent a cardiac catheterization or noninvasive cardiac imaging like CT angiography and cardiac MRI as a standard part of the workup and recommendation for anatomical and functional cardiac evaluation because of limitation of right heart assessment on echocardiography in adult patient.[15-17] In our experience a transthoracic echocardiogram provided sufficient information needed and no patient in our study underwent an invasive or noninvasive imaging modality other than echocardiography. The most probable explanation for this finding in our study is dedicated pediatric hospital who is dealing primarily with pediatric age patient and secondly there is remarkable improvement in echocardiography modality with 2-D imaging with color Doppler, and use of contrast study and wide spread use of trans esophageal echocardiography has nearly preclude need for invasive and noninvasive imaging modalities in double chambered right ventricular obstruction in pediatric patient.[18] Echocardiography provides sufficient preoperative information about anatomical features and associated lesion in double chambered right ventricle. For hemodynamic measurement cardiac catheterization remains gold standard.[19] None of patients in our study required hemodynamics assessment preoperatively.

In our study we have one patient (2%) each of patent ductus arteriosus, Coarctation of aorta, mitral regurgitation and supra mitral membrane. These finding are globally validated as well in different studies as reported by Yuan as Coarctation of aorta 1.6%, pulmonary stenosis 3% and subaortic stenosis 9-35% and said et al who reported an association of pulmonary stenosis 11%, subaortic stenosis 15-36%, right coronary cusp prolapse and aortic regurgitation 15% and double outlet right ventricle in 8%. There were four patients (8%) with pulmonary stenosis, one in isolation and three with ventricular septal defect. We were unable to document any association of atrial septal defect and subaortic stenosis in our study which was well documented in previous published literature as mentioned above.[5,6] The subaortic stenosis is also reported to develop in about 3 % of patient after surgical repair of DCRV on follow up. As our current follow up duration was quite less; we might see patient with subaortic stenosis on long term follow up. Two patient (4%) were having double chambered right ventricle in double outlet right ventricle setting.

The ventricular Septal defect is the most commonly associated abnormality in our study, amounting to 89%. Two previous studies reported ventricular septal defect in 63 to 90% patient of double chamber right ventricle.[20,21] The location of ventricular septal defect was perimembranous sub aortic type in forty (40) patients and six (6) patient have doubly committed sub-arterial type of Ventricular septal defect which is 87% and 13% respectively in our study. The Association of supracristal ventricular septal defect in our study in pediatric patient is higher than previous reported incidence of five percent (5%).[22]

CONCLUSION

In conclusion DCRV was most associated with a VSD of varying size. The association of right coronary cusp prolapse with mid-cavity RVOTO was high in our study along with lack of patient with subaortic stenosis. An early intervention in double chambered right ventricle is associated with low surgical mortality with excellent short- and long-term benefits of prevention of right ventricular dysfunction, low risk of arrhythmia and other risk

associated with excessive ventricular hypertrophy along with decreased risk of aortic cusp prolapse and resulting aortic valve insufficiency.

Conflict of Interest: None.

Authors Contribution

Following authors have made substantial contributions to the manuscript as under:

MR & SAS: Data acquisition, critical review, approval of the final version to be published.

SA & MAK: Data acquisition, data analysis, data interpretation, critical review, approval of the final version to be published.

TK & UK: Conception, study design, data acquisition, drafting the manuscript, approval of the final version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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