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Original Research Article

Clinical, hemodynamic, echocardiographic, angiographic profiles and post-operative outcomes among DCRV patients from a tertiary care referral center in India

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ABSTRACT

Background: This retrospective study of data from 2006 to 2018 at a tertiary care referral center in India aims to document the contemporary clinical and hemodynamic profile of patients who were diagnosed with double chamber right ventricle (DCRV) based on echocardiography and cardiac catheterization. Patients were followed up and their outcomes were studied to document their short and long term outcomes.
Methods: It is a retrospective observational study of patients diagnosed with DCRV in a tertiary care hospital in India. The diagnosis of DCRV was based on the following criteria: an echocardiographic diagnosis of DCRV with doppler evidence of a mid ventricular gradient; cardiac catheterization revealing a systolic pressure gradient between right ventricular inflow and outflow tracts; a right ventricular angiogram demonstrating an anomalous muscle bundle causing obstruction well below the infundibulum. All the patients were followed up for their long term outcomes.
Results: All the patients underwent echocardiography and cardiac catheterization for confirmation of the diagnosis. Sixty percent of our patients presented during adulthood, which is very unusual presentation of this disease entity. Median age of our patient cohort was 23.5 years. Patients presenting during adulthood have atypical symptoms. Dyspnea was the most common presenting symptom in this study. Right ventricular hypertrophy (73.3%)and right bundle branch pattern (26.6%) were the common electrocardiographic findings in our patients. DCRV is commonly associated with other anomalies. Ventricular septal defect (VSD) was the commonest associated anomaly, which was seen in 80% of our patients. Mean gradient cross the anomalous muscle bundle was 67.5 mmHg. Three of our patients (20%) had no associated anomaly, which is very rare in DCRV. Eight patients underwent surgical correction with significant reduction in gradients in all and no perioperative mortality. Median follow up of 8 years showed no adverse outcomes and no progression of gradients.
Conclusions: This study describes in detail the clinical profile, echocardiographic and angiographic identification of anomalous muscle bundles in DCRV patients, which will help the young readers in identifying this often missed diagnosis. It highlights the unusual presentation during adulthood with atypical symptoms in DCRV patients with excellent long-term outcomes on follow up.

Keywords: Adult congenital heart disease, Dual chamber right ventricle, Ventricular septal defect

INTRODUCTION

Double-chambered right ventricle (DCRV) is a rare congenital cardiac disease characterized by anomalous muscle bundles that divide the trabecular right ventricle into two chambers, a high pressure proximal chamber and a low-pressure distal chamber. Right ventricular outflow obstruction is thought to develop progressively over the substrate of a congenital defect.[1-3] This condition is almost always associated with other cardiac defects with

ventricular septal defect (VSD) being the most common, which is present in up to 90% of patients with DCRV.[4] Most cases of DCRV are diagnosed and treated during childhood but infrequently, DCRV can present during adulthood with unusual symptoms causing difficulty in diagnosis.[5] Furthermore, there is a tendency for the obstruction of the right ventricular outflow tract to progress if not treated adequately. Treatment during adulthood is rare and long term data in adults with DCRV is limited.[6]

The aim of this study is to describe the clinical, hemodynamic and angiographic characteristics of pediatric and adult patients with DCRV and their long term surgical outcomes from a tertiary care center from South India.

METHODS

It is a study of all comer population diagnosed to be having DCRV. A retrospective review of records was done in patients with a clinical diagnosis of DCRV from 2008 to 2018 in the Department of cardiology, Christian Medical College-Vellore, which is a tertiary care referral center in India. The diagnosis of DCRV was based on the following criteria: an echocardiographic diagnosis of DCRV with Doppler evidence of a mid ventricular gradient; cardiac catheterization revealing a systolic pressure gradient between the right ventricular inflow and outflow tracts; a right ventricular angiogram demonstrating an anomalous muscle bundle causing obstruction well below the infundibulum. Surgical findings were collected by review of operative notes.

Inclusion criteria included all the patients diagnosed with DCRV based on above criteria.

Exclusion criteria were lack of consent from the patients or inability to verify the diagnosis on both ECHO and cardiac catheterization study.

Fifteen patients with DCRV were identified and their clinical profile, associated cardiac anomalies and hemodynamic parameters on echocardiography and cardiac catheterization were recorded. All the patients were followed up with a detailed clinical examination, electrocardiography, and a complete echocardiographic study. Follow-up data was available varying from 1 to 13 years (median-8 years). Descriptive data for continuous variables are presented as mean or the median and range, as appropriate.

RESULTS

Clinical characteristics

There were 6 females and 9 males. The age of the patients ranged between 2 to 35 years. Nine patients were diagnosed during adulthood, which is a rare presentation. Twelve out of the fifteen patients were symptomatic at presentation while 3 were incidentally detected due to the presence of a murmur. Dyspnea was the most common presenting symptom, seen in 11 cases (73.3%). All patients had audible murmurs in the left parasternal border. There were no patients with cyanosis.

Table 1: Baseline characteristics.

Age	Gender	Presentation	Murmur	Associated anomalies	Treatment
2	F	Recurrent LRTIs	+	Non restrictive VSD	ICR
5	M	Asymptomatic*	+	Restrictive VSD	Medical follow up
5	M	Asymptomatic*	+	Restrictive VSD	Medical follow up
10	F	Dyspnea	+	Restrictive VSD	Medical follow up
11	M	Dyspnea	+	Restrictive VSD	ICR
11	F	Chest pain, dyspnea	+	Restrictive VSD	ICR
18	F	Asymptomatic*	+	Restrictive VSD	Medical follow up
19	M	Dyspnea	+	Restrictive VSD	ICR
28	M	Dyspnea, presyncope	+	Non restrictive VSD	ICR
29	M	Dyspnea	+	Non restrictive VSD	ICR
29	F	Dyspnea	+	None	ICR
29	M	Chest Pain	+	Restrictive VSD	Advised ICR- Not done
33	M	Dyspnea	+	None	Advised ICR- Not done
33	F	Dyspnea	+	None	ICR
35	M	Dyspnea	+	Restrictive VSD	Advised ICR- Not done

*- These patients were identified because of the murmur, VSD- Ventricular septal defect, ICR- Intra cardiac repair, LRTI- Lower respiratory tract infection

Associated anomalies

VSD was the commonest associated anomaly observed in 80% of our cohort. Nine of them had restrictive VSDs while three had non-restrictive VSD. One patient had associated patent foramen ovale (PFO). One patient had coexisting valvular pulmonary stenosis and one had

severe tricuspid regurgitation. Three patients (20%) had no associated anomalies.

Electrocardiographic (ECG) findings

Eleven patients (73.33%) had right ventricular hypertrophy out of which four also had right bundle branch block (RBBB) pattern. Four patients had normal ECG findings.

Hemodynamic findings

The gradient across the anomalous muscle bundle ranged from 12-141 mmHg on echocardiography. On cardiac catheterization, it varied from 14-164 mmHg.

Table 2: Hemodynamic data.

ECHO gdt	RVIP	RVOP	mPAP	Cath gdt	PO gdt
89	89	29	29	40	12
12	42	28	28	14	-
45	55	25	14	30	-
40	48	17	20	31	-
43	80	27	29	53	6
101	62	16	18	46	7
88	104	20	15	84	-
80	145	23	15	122	7
40	83	30	19	53	14
141	88	35	13	53	8
118	164	21	18	143	9
87	194	30	12	164	-
100	111	9	9	56	-
101	123	34	14	89	12
103	136	25	18	111	-

ECHO gdt-Echocardiographic gradient, RVIP-Right ventricular inflow pressure, RVOP-Right ventricular outflow pressure, mPAP-mean pulmonary artery pressure, Cath gdt-cardiac catheterization gradient, PO gdt- post operative gradient

Surgical outcomes

Eight patients (53.33%) underwent surgical intracardiac repair. Seven of them had uneventful postoperative recovery while one patient developed junctional rhythm postoperatively which spontaneously reverted back to sinus rhythm on third post-operative day. The post-operative gradients on echocardiogram were less than 20mmHg in all of them. There was no perioperative mortality. Long term follow up (4-11 years) showed no late mortality or arrhythmia. Patients with mild gradient who were managed conservatively did not show significant worsening of gradients on follow up (longest 13 years).

DISCUSSION

Peacock first described an anomalous obstructive muscle band within the right ventricle in 1867 and Lucas et al described the hemodynamic details in 1962.[2,7] The muscle band divides the trabecular portion of right ventricle into two chambers. It forms a broad triangular band with apex towards crista suprventricularis and base toward the interventricular septum.[8] Superior and rightward displacement of the septal insertion of the moderator band at birth, especially with associated VSD, is thought to be a precursor of DCRV. The degree of superior displacement of the moderator band may help to identify those infants at risk for the development of subpulmonary obstruction.[1] Flow disturbance in the right ventricular outflow tract (RVOT) can cause abnormal proliferation of septoparietal trabeculae and hypertrophy of the displaced moderator band, resulting in a chambered right ventricle.[9] This might explain the frequent association of this anomaly with VSD.

Median age at presentation in our study cohort was 23.5 years, which is higher than the mean age of 18.1 years in study of 48 patients reported by Singh et al.[8] Most of the patients are identified and treated during childhood and presentation during adulthood is very rare.[5] This study highlights 9 cases presenting during adulthood. Eighty percent of our patients were symptomatic at presentation with dyspnea being the most common symptom (73.3%) while Singh et al reported 37.5% of their patients to be asymptomatic and 25% presenting with cyanosis.[8] There was no patient with cyanosis in our cohort. Dyspnea perception could be explained by right ventricular obstruction causing limitation of cardiac output and right ventricular dysfunction may present as fatigue and reduction in exercise capacity.

Eighty percent of our patients had VSD, which is consistent with multiple earlier studies showing VSD being the most common association with DCRV.[4,8] Three patients in our study had DCRV with intact interventricular septum (IVS) and no other associated anomaly. Such a presentation is extremely rare. Small restrictive VSD may close spontaneously with progression of obstruction across the DCRV, which may cause the presentation with worsening dyspnea during adulthood in patients with DCRV and intact IVS.[10] In contrast to the findings in study by Singh et al, only one of our patients had co-existing valvular pulmonary stenosis, which was the second most common (38%) association in their study.[8] There was no patient with cyanotic heart disease in our cohort while Singh et al had 10% cases with double outlet right ventricle.[8] Kottayil et al reported 14 adults with DCRV and all of whom were acyanotic.[10] Right ventricular hypertrophy was the commonest ECG abnormality in patients with DCRV in our study (73.3%) which is similar to that noted by Singh et al who found it in 96%of their patients.[8]

A preoperative diagnosis of DCRV is of paramount importance because anatomical relationships may be difficult to define at operation. A missed DCRV preoperatively can lead to serious adverse outcomes.[7] Failure to identify and correct a DCRV in a patient with a

VSD may lead to a redo surgery at a later date. In patients with intact IVS and those with non-restrictive VSD, the flow acceleration in the right ventricular outflow due to anomalous muscle bundle can be easily identified on echocardiography (Figure 1). However, it may be difficult to detect the anomalous muscles in right ventricular cavity in adult patients with poor anterior resolution and in those with severe right ventricular outflow obstruction. Co-existing VSD or pulmonary stenosis may also be missed due to turbulence in the right ventricular outflow tract (RVOT).[11]

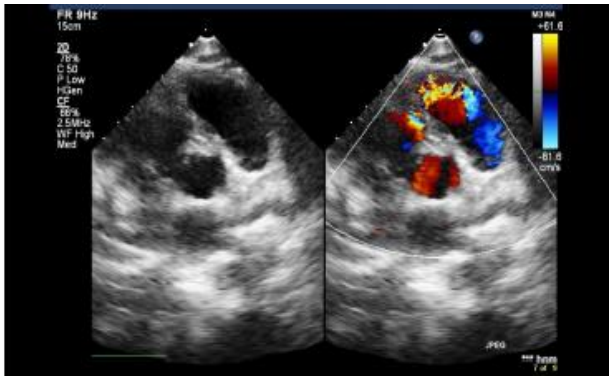

Figure 1: PSAX view at aortic level showing anomalous muscle bundle in the RV outflow with turbulent flow across it.

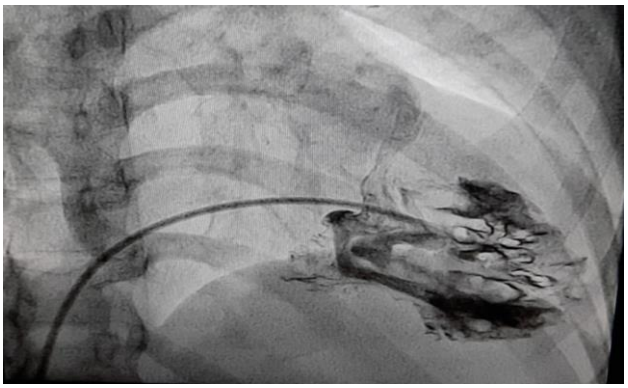

Figure 2: RV angiogram in RAO view (systolic frame) showing anomalous muscle bundle between trabecular inflow and smooth walled right ventricular outflow.

All our patients underwent a cardiac catheterization study which helps in delineating the anomalous muscle bundle, associated anomalies and gradient across the muscle bundle. There was significant difference in the gradient measured on transthoracic echocardiography and on cardiac catheterization, which could be due to difficulty in separating the doppler jet of a restrictive VSD and flow across the anomalous muscle bundle, causing overestimation of gradient on echocardiography. RVOT gradient should be taken from various views (especially subcostal sagittal and coronal views in children and parasternal short axis view at the aortic level in adults) to separate VSD jet from the RVOT. Tricuspid peak velocity can be used to assess RV pressures. Cardiac catheterization delineates the anomalous muscle band in the form of a filling defect which divides the coarsely trabeculated proximal chamber from a relatively smooth walled distal chamber (Figure 2 and 3). We noted that the anomalous muscle bundles are best observed in the antero-posterior view in systolic frame as reported in earlier studies.[8] Right anterior oblique (RAO) view with cranial angulation is another useful view to delineate these anomalous muscle bundles.

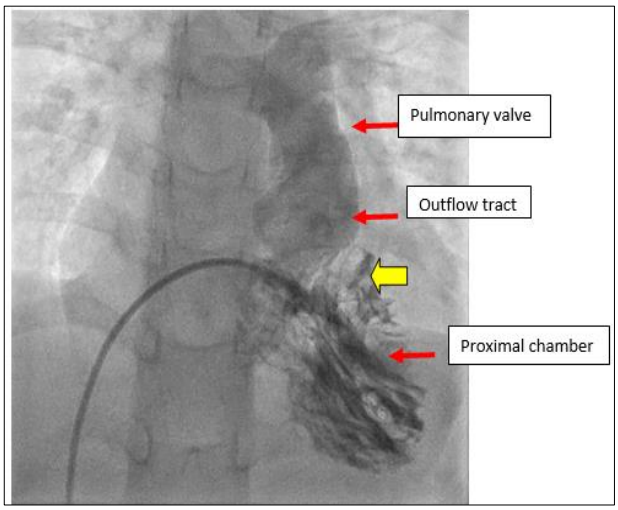

Figure 3: RV angiogram in PA cranial view showing anomalous muscle bundles (yellow arrow) between proximal trabeculated chamber and smooth walled outflow tract.

Treatment of DCRV cases is surgical which consists of the resection of the anomalous muscular bundle and correction of the associated cardiac anomalies. If there are no co existing significant defects, observation is possible as long as the intracavitary systolic gradient less than 40mmHg and the obstruction is not progressive.[5] Eight patients in this series underwent surgical correction majority of which were through trans atrial route. Post-operative outcomes were excellent on short and long term follow up (longest follow up 11 years). There is a paucity of data on long term post-surgical outcomes in adults with DCRV. Five adults in our study who underwent surgical correction had uneventful perioperative period and long term follow up showed an uncomplicated course.

The limitations of this study are that it was retrospective study with a small sample size. Nevertheless, considering the rarity of the disease and presentation, we believe that this study provides important clinical understanding to DCRV. We acknowledge that a longer follow up is pertinent to detect the development of late arrhythmias and progression of obstruction in those with anomalous muscle bundles and low gradients.

The key highlights of this study are as follows:

- There is a paucity of data on adult DCRV with their long term post-surgical outcomes as most are identified and treated during childhood. This is one of the few studies in literature which highlights the clinical, echocardiographic and angiographic profile of this subset of patients and their long term post-surgical outcomes.
- A high index of suspicion of DCRV has to be kept in mind while assessing a patient with VSD with flow turbulence in right ventricle, as failure to identify and correct DCRV in patients with VSD may lead to redo surgery.
- This study with a small sample size highlights the need of further studies with bigger sample size to evaluate long term outcomes of adults with DCRV to study for late complications and disease progression.

CONCLUSION

This series provides the contemporary data on clinical and hemodynamic profiles of a rare disease from a tertiary care referral center in India. As compared to earlier reported studies, this study highlights the atypical presentation during adulthood with atypical symptoms in most of our patients. Twenty percent of our patients had DCRV with no associated anomaly which is very unusual.

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Ethical approval: The study was approved by the Institutional Ethics Committee

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