Double-chambered Right Ventricle in Children

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ABSTRACT

A double-chambered right ventricle (DCRV) is a rare congenital heart defect. The clinical features of DCRV, especially in the pediatric population, have not been sufficiently elaborated. There are many unanswered questions regarding the surgical indications, surgical timing, natural history, and patient outcomes. This article will discuss the definition, pathophysiology, clinical features and treatment of DCRV. This systematic review showed a higher prevalence of symptomatic patients among children with DCRV with signs of right ventricular pressure overload and associated congenital heart defects. The spatial relation between ventricular septal defect and the anomalous muscle bundle might be a determinant of right ventricular obstruction. Symptomatic patients with an intra-right ventricular pressure gradient of >20 mmHg are indicated for a surgical repair. DCRV has a favourable postoperative course as no postoperative death was reported. Mechanisms of right ventricular obstruction warrant further elucidations based on larger patient population in the near future.

Key Words: Cardiac surgical procedures, Heart defects, Congenital, Heart ventricles.

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INTRODUCTION

A double-chambered right ventricle (DCRV) is a rare congenital condition in which the right ventricle is divided into a proximal high-pressure chamber and a distal low-pressure chamber by abnormal muscle bundles.¹ In such patients, right ventricular outflow tract obstruction often develops especially in those with an abnormally short distance between the moderator bundle and pulmonary valve.² The presence of abnormal muscle bundles can produce a pressure gradient between the inflow and outflow of the right ventricle.³ The obstruction can be progressive with time.⁴ Debate persists regarding the exact pathogenesis of the obstructing muscle bundles, although it was proposed that the mechanisms were hypertrophy of the moderator band arising abnormally high from the interventricular septum, and hypertrophy or displacement of the accessory septo-parietal/septomarginal trabeculations.⁴

An isolated DCRV is very rare, representing only 6.2% of the patients, while approximately 80-90% of the DCRV cases are associated with various congenital heart defects, with ventricular septal defect (VSD), in particular a perimembranous type VSD, being the most common. According to Hoffman, the most frequent associated congenital heart defect in DCRV patients

was VSD, which accounted for 84.4%, followed by membranous subaortic stenosis (31.3%).⁵ DCRV can also be an associated anomaly of Williams syndrome.⁶ This anomaly is often diagnosed during childhood and adolescence, while very few were found in adults.⁷ However, there is no consensus on surgical indications and on the optimal timing of surgical operation in terms of the severity of obstruction and the hemodynamic burden of the right ventricle.⁴

The aim of this review was to discuss the definition, pathophysiology, clinical features, and treatment of pediatric DCRV.

METHODOLOGY

The Preferred Reporting Items for Systematic Reviews and Meta-analyses (PRISMA) statement guidelines were followed in this meta-analysis. Publications were systematically searched in the PubMed, Highwire Press, and the Cochrane Library databases from January 2000 to December 2018. The MeSH terms and keywords used to identify articles included "double-chambered", "two-chambered", "right ventricle", "double chambered right ventricle (DCRV)", "anomalous muscle bundles", and "hypertrophied moderator band", with a filter of "pediatrics". The screening of the bibliographic references helped in completing the literature retrieval. Forty-two articles were found related to the topic and keywords in the literature search; and 18 articles, which met the inclusion and exclusion criteria during preliminary assessment, were included in the review. The exclusion criteria were articles of double-chambered left ventricle (n=7), DCRV of adult patients (n=5), no substantial information of pediatric DCRV patients (n=4), pediatric

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patient information unable to be sorted out from that of adult patients (n=2), pediatric patient with a previous history of DCRV repair (n=1), and being irrelevant to pediatric DCRV (n=5).

The data independently extracted from each study were the study population, demographics, clinical manifestations, diagnostic techniques, pre- and postoperative intra-right ventricular pressure gradients, associated disorders, conditions of the infundibulum, anomalous muscle bundles, therapeutic strategies, and patient outcomes.

The measurement data were expressed in mean $\pm$ standard deviation, and were compared by independent sample/paired t-test. The categorical variables were presented as frequency and percentage, and were compared by Fisher exact test. $p < 0.05$ was considered statistically significant. An IBM SPSS statistics version 22.0 was used to perform the statistical analyses.

RESULTS

In total, 18 articles reporting pediatric DCRV were collected,^{4,8-24} with 3 original articles,^{4,8,15} 1 case series;¹⁸ and 14 case reports included.^{9-14,16,17,19-24} As a result, as many as 62 pediatric patients were recruited in this study. One of the patients was a fetus, who was diagnosed with DCRV and also delivered at 28-week gestation, and died at the age of 6 hours. This case was approximately regarded as at an age of 0. Thus, patients of this setting were at a mean age of 10.4 ± 5.2 years (n=18). The gender of 38 patients was known: 18 (47.4%) were males and 20 (52.6%) were females ($\chi^2=0.2$, $p=0.819$). There was no age difference between male and female patients (13.0 ± 3.7 years vs. 11.4 ± 5.8 years, $p=0.551$).

Clinical manifestations were described for 20 (20/62, 32.3%) patients, who had 28 symptoms. Three (10.7%) patients were asymptomatic, and 25 (89.3%) were symptomatic ($\chi^2=34.6$, $p<0.001$). Of the symptomatic patients, dyspnea/shortness of breath was the most common presenting symptoms (Table I).

A heart murmur was audible in 10 (10/62, 16.1%) patients. In one patient, the heart murmur was unspecified.¹⁶ Of the remaining 9 patients, all had one or more systolic murmurs. The heart murmur was audible at the left sternal border in 4 (44.4%) patients,^{9,13,14,17} at the right sternal border in 3 (33.3%) patients,^{10,22,24} and at precordium²⁰ and at all foci¹² in 1 (11.1%) patient, respectively.

Among 6 patients with an electrocardiogram examined, 2 (33.3%) patients had sinus tachycardia,^{9,13} 1 (16.7%) patient had normal sinus rhythm,²² and 3 (50%) patients showed signs of left/right ventricular hypertrophy or left atrial enlargement.^{10,12,14}

The locations/natures of the hypertrophied anomalous muscle bands were heterogeneously described for 30 patients (Table II).

An intra-right ventricle pressure gradient was measured by echocardiography in 51 (51/62, 82.3%) patients,^{4,8-17,20,23,24} and by catheterisation in 27 (27/62, 43.5%) patients,^{4,8,15} with a mean pressure gradient of 92.8 ± 27.6 (rang, 45-141; median, 86) mmHg (n=17).

The infundibulum of the right ventricle was described in 6 patients: it was normal in 2 (33.3%) patients,^{13,14} and

Table I: Clinical symptoms.

Symptom	n (%)
Asymptomatic ^{20,22,24}	3 (10.7)
Symptomatic	25 (89.3)
Dyspnea/shortness of breath ^{4,11,16,17,23}	9 (36)
Cyanosis ^{4,10}	2 (8)
Palpitation ^{9,12}	2 (8)
Heart failure ^{9,13}	2 (8)
Fatigue ^{12,17}	2 (8)
Cardiac arrest ²¹	1 (4)
Exercise intolerance ⁴	1 (4)
Easy fatigability ⁹	1 (4)
Loss of appetite ⁹	1 (4)
Fever ⁹	1 (4)
Growth retardation ¹⁴	1 (4)
Hydrops fetalis ¹⁹	1 (4)
Weight loss ^{6,9}	1 (4)

Table II: The locations / natures of the hypertrophied anomalous muscle bands.

Locations/natures of anomalous muscle bands	n (%)
Ventricular septal defect was proximal to the obstructing muscle bundle ¹⁵	(44.8) 13
Distal to the bundle ¹⁵	8 (27.6)
In the right ventricular outflow tract region ^{9,17}	2 (6.9)
At the infundibular and right ventricular outflow tract ¹¹	1 (3.4)
At the infundibular region ¹²	1 (3.4)
Hypertrophied moderator band ¹⁰	1 (3.4)
Between the septal tricuspid valve and the anterior wall of right ventricle ¹³	1 (3.4)
In the roof of the right ventricular chamber ¹⁴	1 (3.4)
Between the thin-walled infundibulum and the inflow segment of the right ventricle ¹⁶	1 (3.4)

Table III: 64 associated congenital cardiac disorders in 53 patients.

Associated congenital cardiac disorders	n (%)
VSD	49 (76.6)
VSD, perimembranous ^{4,9,11,15,18,23,24}	34 (69.4)
VSD, muscular ^{4,15}	6 (12.2)
VSD, multiple, apical ²¹	1 (2.0)
VSD, supracristal ¹⁵	1 (2.0)
VSD, unspecified ⁸	7 (14.3)
Subaortic aortic stenosis ^{4,8,17}	6 (9.4)
Atrial septal defect ^{8,10}	2 (3.1)
Pulmonary stenosis ⁴	2 (3.1)
Aneurysm of the membranous interventricular septum ²²	1 (1.6)
Aortic coactation ⁴	1 (1.6)
Asymmetric septal hypertrophy (IVS 2.85 cm, with no LVOTO) ¹²	1 (1.6)
Dextracardia ¹⁰	1 (1.6)
Right aortic arch ⁴	1 (1.6)

IVS = Interventricular septum; LVOTO = Left ventricular outflow tract obstruction; VSD = Ventricular septal defect.

abnormal in 4 (66.7%) patients ($\chi^2=1.3$, $p=0.567$) [including a hypertrophied anomalous muscle at the infundibular region ($n=2$),^{11,12} a narrow infundibulum due to hypertrophy ($n=2$),¹⁰ and a thin-walled infundibulum ($n=1$).¹⁶

Ventricular hypertrophy was seen in 12 (12/62, 19.4%) patients: it was right ventricular hypertrophy in 10 (83.3%) patients, biventricular hypertrophy in 1 (8.3%) patient, and biventricular hypertrophy with right atrial enlargement in 1 (8.3%) patient, respectively ($\chi^2=20.3$, $p<0.001$).

Nine (14.5%) patients had an isolated DCRV,^{8,13-16,18-20} and in 53 (85.5%) patients, the DCRV was associated with one or more congenital heart defects ($\chi^2=62.5$, $p<0.001$).

The latter 53 patients had 64 associated congenital heart defects, with VSD being the most common (Table III). Of the patients with a VSD, the perimembranous type VSD was the most common, accounting for 81.0% (34/42) by excluding the cases with an unspecified VSD type.

Three (4.8%) patients were complicated with infective endocarditis,^{4,13} and 1 (1.6%) patient presented with multiple vertebral malformations, mental retardation, microcephaly, facial dysmorphism and growth retardation.²²

The treatment of choice was reported for 20 (32.3%) patients: 19 (95%) patients were surgically treated, and 1 (5%) patient was conservatively managed ($\chi^2=32.4$, $p<0.001$).

Of the 19 surgically treated patients, anomalous muscle bundle resection was performed in all cases. Besides, 5 (5/19, 26.3%) patients had one or two concurrent intracardiac operations, including pulmonary valve vegetation excision,¹³ VSD closure,⁹ VSD closure and right ventricular outflow tract enlargement,²⁴ pulmonary valvotomy and VSD closure,¹¹ and subaortic membrane resection and VSD closure¹⁷ in one patient each. The surgical incisions were indicated in 15 (15/19, 78.9%) patients: 13 (86.7%) patients were *via* a right ventriculotomy and 2 (13.3%) were *via* a right atriotomy ($\chi^2=16.1$, $p<0.001$).

Ozeke *et al.*²² reported managing their asymptomatic DCRV patient conservatively, but they did not specify the therapeutic agents.

The outcomes of 34 (54.8%) patients were reported: 27 (79.4%) patients were event-free survivors, 6 (17.6%) patients had complications, and 1 (2.9%) patient died. Of the 6 patients with complications, 4 (66.7%) patients had postoperative complete atrioventricular block and 2 (50%) of them required permanent pacemaker implant.^{4,8} One (16.7%) patient had subacute constrictive pericarditis, which was cured by partial pericardiectomy and a 2-week prednisolone administration,²⁴ and 1 (16.7%) patient developed postoperative transient blindness,

which was significantly improved after conservative treatment.¹⁰ There was no postoperative death.

The asymptomatic patient with DCRV can be conservatively managed and followed up as indicated by Ozeke *et al.*²²; however, they did not state the length of their follow-up on their patient. Thirteen (21.0%) patients were on a postoperative follow-up ranging from 1-192 months.^{4,8,11,12} Postoperative intra-right ventricular pressure gradient was reported for 17 patients. It was detected by echocardiography in 11 patients,^{8,12,14,16} and the detection method was not specified for the remaining patients. The pressure gradient was significantly lower than the preoperative value (92.8 ± 27.6 mmHg vs. 3.6 ± 5.6 mmHg, $p<0.001$).

DISCUSSION

The etiology of obstruction was anomalous muscle bundles across the right ventricular cavity in 41 patients (67%), right ventricular midventricular fibrous diaphragm in 19 (31%) patients, hypertrophied moderator band in 5 (8%) patients, and hypertrophied papillary muscle of the conus in 1 (1.6%) patient.²⁵ Three levels of obstruction were present: low in 37 (61%) patients, high in 14 (23%) patients, and mixed level in 10 (16%) patients.²⁵ The increased blood flow and pressure within the right ventricular outflow tract may act as an initial stimulus for hypertrophy of the *crista supraventricularis* in patients with VSD and notably increased pulmonary blood flow.⁵

Research on spatial relations between VSD and anomalous muscle band revealed that the VSD was proximal to the obstructing muscle bundle in 13 patients (62%), and distal to the bundle in 8 (38%).¹⁵ The pressure gradient within the right ventricle averaged 67 mmHg in the proximal group and 83 mmHg in the distal group.¹⁵ However, in literature, the relation between VSD and muscle band was not indicated in most instances. In some series, the VSD was noted to open to the proximal chamber in all cases; while in others, it opened to the distal chamber thus acting as an extension of the left ventricle.¹⁵ In general, VSD was proximal to the anomalous muscle band in 2/3 cases.¹⁵ The severity of right ventricular obstruction is progressive in some unoperated patients, with a speed of progression of 1.5-6 mmHg/year.⁴ Progression of right ventricular obstruction can be due to muscle hypertrophy and/or endocardial fibrosis.⁴ No patient required reoperation for recurrence of intra-right ventricular obstruction, and no patient died during adulthood.⁴ The average peak pressure gradient across the VSD was 31 mmHg when the VSD was proximal and 54 mmHg when it was distal to the obstructing bundle.¹⁵

Obstructions of DCRV can be divided into three types: low and oblique obstruction, high and horizontal obstruction, and mixed obstruction.⁵ An angiographic study in 10 DCRV patients revealed an oblique and low

obstruction in 6 patients and a high and horizontal obstruction in 4 patients.⁸ Hypertrophic septal and parietal muscle bundles might cause infundibular stenosis.¹³

Alva *et al.*⁸ proposed certain criteria of diagnosis and surgical indications of DCRV. These include an intra-right ventricular pressure gradient >20 mmHg by echocardiography or by cardiac catheterisation; an angiographic evidence of intra-right ventricular obstruction; normal right ventricular infundibulum; and surgical confirmation of DCRV during the operation. However, the criterium of "normal right ventricular infundibulum" could be suspicious, as it could be hypertrophied, narrowed, or thin-walled in DCRV patients, as shown in this report.

The presenting symptoms of patients with DCRV include syncope, angina, and severe dyspnea; clinically resembling pulmonary artery hypertension,² and the high right ventricular systolic pressure is sometimes falsely interpreted as pulmonary artery hypertension.²⁶ Therefore, DCRV should be firstly differentiated with pulmonary artery hypertension.²⁷

DCRV can be misdiagnosed as VSD and lead to mis-treatment.²⁸ The mid-cavitary turbulent jet on transthoracic echocardiography can be mistaken as an intracardiac shunt, and thus, caution should be taken to differentiate DCRV from isolated VSD.²⁶ The subcostal view of transthoracic echocardiography (TTE) may sometimes provide better visualisation of the right ventricle and right ventricular outflow tract obstruction.²⁶ Moreover, it was once reported that 2 patients were reported development of DCRV 6-8 years after the surgical closure of VSD.²⁸

Anomalous muscle bundles in DCRV may cause right ventricular outflow tract obstruction.²⁹ Failure to detect the anomalous muscles in right ventricular cavity may lead to a misdiagnosis of severe right ventricular outflow obstruction and myocardial hypertrophy. Thus, it should be differentiated with other congenital heart defects causing right ventricular outflow tract obstruction, such as tetralogy of Fallot. Two dimensional echocardiography with Doppler flow analysis is useful in the evaluation and differential diagnosis of right ventricular outflow obstructions prior to invasive studies and surgical intervention.³⁰

Kahr *et al.*⁴ reported the follow-up results of 50 patients with DCRV. They found 47% of the patients were asymptomatic despite significant right ventricular obstruction, and 16% of these were completely asymptomatic during follow-up with conservative management. The remaining 42 (84%) patients became symptomatic at a median age of 26 years. Out of these, 33 (78.6%) patients were operated on and 91% of the operated patients showed no residual intra-ventricular gradient, and 27/33 of the patients had improved cardiac function. There was no

early or late mortality and no re-intervention requirement for DCRV during a median follow-up of 8 years.

It has been suggested that the surgical resection of the anomalous muscle bundles should be performed once the diagnosis of DCRV is made.⁴ However, this approach is not popularly accepted. When the patient becomes symptomatic, or with progressive right ventricular outflow stenosis or progressive right ventricular inflow pressure overload, an early surgical repair is then warranted. Nevertheless, debates remain concerning the surgical indications for asymptomatic DCRV patients.¹ It has been proposed that right ventricular outflow tract obstruction with a pressure gradient of 20-50 mmHg, detected by echocardiography or by cardiac catheterisation, is an indication for DCRV repair.⁴ Other authors have recommended close follow-up for patients with mild non-progressive intraventricular gradient without associated cardiac defects.²² As it has been observed that sustained monomorphic ventricular tachycardia in patients with unoperated DCRV was resolved after surgical removal of the obstructing muscular bundles; therefore, a surgical treatment of DCRV is indicated for the patients with evidence of ventricular tachycardia.⁴

Kahr *et al.*⁴ reported that DCRV repair was carried out *via* a median sternotomy in all their patients, in whom a right ventriculotomy was used in 36% patients, a combined right atriotomy and right ventriculotomy in 32%, a pulmonary arteriotomy in 23%, and a transatrial approach in 9% of the patients. Right ventriculotomy was generally avoided in patients operated after 2001 due to the concern of right ventricular function depression and ventricular tachycardia secondary to right ventriculotomy.⁴ Nevertheless, in adult patients with advanced obstruction and a high right ventricular outflow pressure gradient, a right ventriculotomy is sometimes warranted for the relief of obstruction.¹

Ozeke *et al.*²² reported briefly to manage their asymptomatic DCRV patient conservatively, but they did not specify the therapeutic drugs. No other authors adopted conservative treatment for DCRV patients. By searching the literature, it was found that none described the use of β -blockade and calcium channel blockade in either pediatric or adult patients with DCRV.

DCRV has a favourable postoperative course and no postoperative death was reported. The likelihood of DCRV recurrence is very low; however, postoperative follow-up was always less than 10 years in most series.¹

CONCLUSION

A DCRV is a rare congenital heart defect. Most of the patients are symptomatic with signs of right ventricular pressure overload and are associated with other congenital heart defects, especially perimembranous VSDs. The spatial relation between VSD and the

anomalous muscle bundle might be a determinant of right ventricular obstruction. Symptomatic patients with an intra-right ventricular pressure gradient of >20 mmHg are indicated for a surgical repair. DCRV has a favourable postoperative course as no postoperative death was reported. Mechanisms of right ventricular obstruction warrant further elucidations based on large patient population in the near future.

ETHICAL APPROVAL:

Ethical approvals were obtained from the Institutional Review Board prior to initiation of the research work.

CONFLICT OF INTEREST:

The author declares no conflict of interest.

AUTHORS' CONTRIBUTION:

SMY: Substantial contribution to the conception and design of the work; and the acquisition, analysis, and interpretation of data for the work; drafting the work and revising it critically for important intellectual content; final approval of the version to be published; agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy and integrity of any part of the work are appropriately investigated and resolved.

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