Double-Chambered Right Ventricle

Diagnosis and Treatment

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ABSTRACT

Background: Double-chambered right ventricle is better comprehended as a type of septated right ventricle caused by abnormally located or hypertrophied muscular bands, which separates the right ventricle cavity into a proximal and a distal chamber. Common related defects incorporate ventricular septal defect, discrete subaortic stenosis and pulmonary valve stenosis.

Objectives: To review presenting features, diagnosis, associated defects and the results of surgical treatment of double-chambered right ventricle.

Methods: Nine patients under went surgical repair of double-chambered right ventricle. Their median age was 16.7 years, five patients were females and four patients were males. Their data were collected and retrospectively studied from 1st of October 2012 to 1st of January 2017 at Ibn-Alnafees teaching hospital for cardiothoracic surgery in Baghdad, Iraq. We reviewed the patient's age, sex, clinical presentations, preoperative variables, diagnostic modalities, surgical approach, intraoperative variables and postoperative morbidity and mortality.

Results: There was no intra-operative death, mortality was 0%. Wound infection was common post-operative complication 66.7%. Exertional dyspnea was common clinical presenting features 33.3%. Ventricular septal defect was common associated anomaly 66.7%. Transthoracic echocardiography was diagnostic modality in all patients 100%. Transatrial approach was common surgical approach 88.9%.

Conclusions: Double-chambered right ventricle usually associated with ventricular septal defect and subaortic stenosis. Surgery of repair double-chambered right ventricle associated with low morbidity and mortality, that's mean double-chambered right ventricle could be considered as a disease with good surgical outcome.

Keywords: **Anomalous muscle bands, Transatrial approach, Ventricular septal defect, Exertional dyspnea.**

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Double-chambered right ventricle (DCRV) is a type of right ventricular (RV) outflow tract obstruction caused by anomalous muscular or fibromuscular bundles that separates the right ventricle into proximal high-pressure and distal low-pressure chambers[1,2]. Male to female proportion is 2:1[3]. These lesions regularly present in pediatrics age groups[4]. About 80% to 90% of patients DCRV is associated with other congenital anomalies[5]. Isolated DCRV is extraordinary[5]. Ventricular septal defects (VSD) is the most associated anomaly, afterward being pulmonary stenosis.

Other relations are tetralogy of Fallot, pulmonary atresia with intact ventricular septum, transposition of the great arteries, double outlet right ventricle, Ebstein anomaly and anomalous pulmonary venous drainage[5].

Presentation is usually in youth however can be as right on time as infant period[6]. The commonest symptoms are dyspnea and diminished exercise resistance. Anyway endocarditis, cyanosis, heart failure, chest pain, and syncope are accounted for[7]. Transthoracic echocardiography (TTE) might be lacking for diagnosis, so transesophageal echocardiography (TEE) is firmly prompted for both youngsters and grown-ups,

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especially within the sight of right ventricular hypertrophy on electrocardiogram[8]. Cardiac catheterization might be performed to affirm the determination[9]. Right ventricular angiography indicating filling defects inside the right ventricle between the outflow and inflow zones, affirms the diagnosis[9]. Left ventriculography is performed for related VSD or subaortic stenosis[9]. As of late, contrast computed tomography and cardiac magnetic resonance have been presented. Such methods supplant the utilization of invasive right-sided cardiac catheterization[10].

Surgery is indicated in presence of symptoms or where the peak gradient exceeds 40 mm Hg in asymptomatic patients[11]. Resection of the anomalous muscle bands can be utilized in different approaches, including right atriotomy, right ventriculotomy, or combined transatrial-transpulmonary access[12]. The right atriotomy and the combined transatrial-transpulmonary incision are generally utilized[13]. Right ventriculotomy is used rarely because of high incidence of RV dysfunction and ventricular arrhythmias[14].

The aim of this study is to review presenting features, diagnosis, associated defects and the results of surgical treatment of double-chambered right ventricle.

Methods

Nine patients ranged from nine to 30 years of age, median age was 16.7 years. Five patients were females and four patients were males, (Table 1). All underwent surgical repair for DCRV. Their data were collected and retrospectively studied from 1st of October 2012 to 1st of January 2017 at Ibn-Alnafees teaching hospital for cardiothoracic surgery in Baghdad, Iraq. All cases were referred from pediatric cardiology unit to cardiac surgery unit and evaluated by a pediatric cardiologist pre- and postoperatively. Electrocardiography, chest x-ray and TTE used for all patients while TEE was not used in any patient. Cardiac catheterization was performed in one patient to qualify concurrent aortic valve incompetence which was grade 4.

The criteria used for the diagnosis of DCRV were as documentation at TTE (the 50% rule, the absence of fibrous discontinuity between the aortic valve and adjacent atrioventricular valve, the morphology of the conus septum). All patients had associated anomalies, five patients had concurrent VSD and subaortic membrane, one patient had concurrent VSD only, one patient had coexisting ostium secundum ASD and one patient had concurrent valvular pulmonary stenosis. Tetralogy of Fallot was concurrent anomaly in one patient and two patients had concurrent aortic valve incompetence.

Surgery was done using classical median sternotomy, total cardiopulmonary bypass and antegrade cardioplegia in all patients. Intraoperative TEE was not employed in any patient. All associated anomalies were repaired and aortic valve replacement done in one patient. All cases assessed postoperatively by transthoracic echocardiography in early postoperative period and before discharge. We reviewed the patients' age, sex, clinical presentations, preoperative variables (age, sex, body weight, body surface area and preoperative mean pressure gradient), diagnostic modalities, surgical approach, intraoperative variables (aortic cross-clamp time cardiopulmonary bypass time), intubation time, intensive care unit stay, need full inotropic support time and postoperative morbidity and mortality.

The statistical package for the social sciences program version 22 was used for authentic examination. The results were imparted by mean and standard deviation for tenacious components or with repeat and rate for total variables.

Table 1: Age of patients.

Age (years)	No.	%
9-10	5	55.6
11-20	1	11.1
21- 30	3	44.4
Total	9	100

Results

Exertional dyspnea was common clinical presenting feature (33.3 %) while fatigue represent (22.2%), (Table 2).The mean value of preoperative mean pressure gradient was 62 mmHg, (Table 3). VSD was common associated anomaly, which was present in six patients (66.6%), subaortic membrane in five cases (55.5 %) while aortic incompetence in two cases (22.2%), (Table 4). TTE was the diagnostic modality in all patients, cardiac catheterization was applied in one patient to qualify associated aortic valve incompetence, (Table 5). Transatrial approach was common surgical approach and was applied in eight patients (88.9%) versus transatrial transpulmonary, which was applied in one patient only (11.1%), transventricular approach was not used, (Table 5).

Six patients underwent concurrent VSD repair while five patients underwent concurrent subaortic membrane resection. Each of concurrent ASD repair, TOF repair, aortic valve replacement and pulmonary valvotomy were used in one patient for each, (Table 5). There was no intra-operative death. Wound infection was common post-operative complications six cases (66.6 %), respiratory tract infection four cases (44.4%). Pericardial effusion after surgery was seen in three cases (33.3%) while postoperative tachyarrhythmia and tricuspid incompetence were found in two cases (22.2%). Postoperative bleeding was present in one case (11.1%). Residual RV stenosis, postoperative acute renal failure, postoperative RV systolic dysfunction and postoperative heart block were not observed in any patient, (Table 6).

Table 2: Main presenting clinical features.

Variable	No.	%
Exertional dyspnea	3	33.3
Fatigue	2	22.2
Arrhythmia	1	11.1
Cyanosis	1	11.1
Chest pain	1	11.1
Asymptomatic (significant echo finding for operation)	1	11.1
Total	9	100

Table 3: **Mean values of preoperative, intraoperative and post-operative variables.**

Pre-operative variable	Mean value
Body weight (kg)	34.9
Body surface area (m^2)	0.87
Preoperative mean pressure gradient	62
Intra-operative variable	
Aortic cross clamp time (min)	48
Cardiopulmonary bypass time (min)	72
Post-operative variable	
Intubation time (hrs)	4.6
Intensive care unit stay (day)	2.3
Postoperative mean pressure gradient	18
Need full inotropic support time (hrs)	0

Table 4: Associated anomalies.

Variable	No.	%
VSD	6	66.6
Subaortic membrane	5	55.5
Aortic incompetence	2	22.2
ASD	1	11.1
TOF	1	11.1
Pulmonary valve stenosis	1	11.1

Table 5: Diagnostic modalities and surgical approach used.

Diagnostic modality	No.	%
TTE	9	100
Cardiac catheterization	1	11.1
Surgical approach		
Transatrial	8	88.9
Transatrial–transpulmonary	1	11.1
Total	9	100
Additional procedure		
VSD repair	6	66.7
ASD repair	1	11.1
Sub aortic resection	5	55.5
TOF repair	1	11.1
Aortic valve replacement	1	11.1
Pulmonary valvotomy	1	11.1

Table 6: Morbidity after surgery.

Variable	No.	%
Pericardial effusion	3	33.3
Tachyarrhythmia	2	22.2
Respiratory tract infection	4	44.4
Wound infection	6	66.7
Bleeding	1	11.1
Tricuspid incompetence	2	22.2

Discussion

Most instances of DCRV end up clear all through youth. The time of presentation normally shifts from 4 months to 20 years[4]. In the present study age group 21-30 years represented the majority (44.4%) which reflect delay in diagnosis in our center than that abroad simply because of delayed patient referring to a specialized center from other non-specialized centers. Male-to-female ratio is 2:1 as describe in international studies while this study show male: female ratio was 1:1.25 which may reflect high percentage of females in our population[3]. In the present study, exertional dyspnea represented (33.3%) of clinical presentation versus fatigue (22.2%) while the incidence of arrhythmia, cyanosis, chest pain and asymptomatic patients were (11.1%) while the study of Cil and colleagues found that 40% were asymptomatic, 35% exhibited fatigue and 17% presented with dyspnea while palpitation, cyanosis and congestive heart failure constituted 10 to 12% of presentation symptoms[7]. This reflects much delay in diagnosis in our center until exertional dyspnea appeared; the majority of cases were asymptomatic in other centers worldwide. In spite of the fact that TTE is a vital method for diagnosing congenital heart disease in children, it didn't indicate DCRV well in grown-up patients because of representation of the RV is constrained by its unpredictable shape and retrosternal location. Previous contemplates have demonstrated that TTE can yield a precise determination of DCRV in just 8 to 17% of grown-up patients. Lascano and colleagues in their investigation demonstrated that TEE gives preferable perception of the RV over does TTE[3], they (Lascano and colleagues) suggested the

utilization of right-sided cardiac catheterization in assessing grown-up patients with suspected RV outflow tract obstruction and VSD[3]. In this study TEE was used to assist in the diagnosis in all age groups in addition to other traditional diagnostic modalities. TEE was not used in any patients (because of its shortage) as that of right-sided cardiac catheterization so there were many missed cases due to under diagnosis of cases because of depending on TTE only for diagnosis. In this study, all cases had associated defects 100%. VSD was common associated defect represent 66.7% while in other study the association with VSD was 80% to 90%, this may be due to small size of the current sample[5]. An outstanding relationship is depicted among patients with RV outflow tract obstruction, VSD, and subaortic stenosis. Vogel et al depicted 36 patients with VSD and double-chambered right ventricle, 88% of whom had echocardiographic proof of subaortic stenosis[5]. In our investigation six patients with DCRV and VSD, 83.3% of them had subaortic stenosis. In present study, there was no operative death, which reflects safety of DCRV repair. Residual RV stenosis was 0% so feasibility of surgery to resect abnormal muscle was excellent. Mean intubation time and intensive care unit stay was short and also there was no need of full inotropic support which reflect smooth postoperative period. Acute renal failure, RV systolic dysfunction and postoperative heart block were not observed in any case. Wound infection, respiratory tract infection, mild pericardial effusion and self-limited tachyarrhythmia were the mean complications which reflect low morbidly of surgery of DCRV. Transatrial approach was used in 889% of cases, transatrial - transpulmonary approach was used in 11.1% depending on the size of defect and surgeon experience and there was no need of transventricular approach which reflect good experience in surgical resection of abnormal muscle in our center without utilizing ventriculotomy which associated with postoperative RV systolic dysfunction and tachyarrhythmia which match that abroad[12]. The study limitation is lack of long-term follow up.

In conclusion, DCRV usually associated with VSD and subaortic stenosis. A more experience of usage TEE and right side catheterization for diagnosis DCRV is needed. Low morbidity and mortality were noticed in the center.

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