Cardiology in the Young

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Early and mid-term outcomes of double-chambered right ventricle repair: An 8-year experience

Vikram Halder[1], Soumitra Ghosh[2], Shyam Kumar Singh Thingnam[3], Harkant Singh[3], Anand Kumar Mishra[3], Sachin Mahajan[3], Rupesh Kumar[3], Pankaj Aggarwal[3], Aduri Raja Suman Dutta[3] and Amit Mishra[1]

[1]Department of CTVS, UNMICRC, Ahmedabad, India; [2]Department of Cardiology, PGIMER, Chandigarh, India and [3]Department of CTVS, PGIMER, Chandigarh, India

Original Article

Cite this article: Halder V, Ghosh S, Thingnam SKS, Singh H, Mishra AK, Mahajan S, Kumar R, Aggarwal P, Dutta ARS, and Mishra A (2024) Early and mid-term outcomes of double-chambered right ventricle repair: An 8-year experience. *Cardiology in the Young* **34**: 268–271. doi: 10.1017/S1047951123001531

Received: 14 April 2023
Revised: 25 May 2023
Accepted: 25 May 2023
First published online: 15 June 2023

Keywords:
Double-chambered right ventricle; ventricular septal defect; surgical repair; surgical outcomes

Correspondence author: V. Halder;
Email: vikramavnrt@gmail.com

Abstract

Background/Aim: Double-chambered right ventricle is a rare and progressive condition that is characterised by obstruction of the right ventricular tract. Double-chambered right ventricle is usually associated with ventricular septal defect. Early surgical intervention is recommended in patients with these defects. Based on this background, the present study aimed to review early and midterm outcomes of primary repair after double-chambered right ventricle. *Methods:* Between January 2014 and June 2021, 64 patients with a mean age of 13.42 ± 12.31 years underwent surgical repair for double-chambered right ventricle. The clinical outcomes of these patients were reviewed and assessed retrospectively. *Results:* An associated ventricular septal defect was present in all the recruited patients; 48 (75%) patients of sub-arterial type, 15 (23.4%) of perimembranous, and 1 (1.6%) patient of muscular type. The patients were followed up for a mean period of 46.73 ± 27.37 months. During their follow-up, a significant decrease in the mean pressure gradient from 62.33 ± 5.52 mmHg preoperatively to 15.73 ± 2.94 mmHg postoperatively was observed ($p < 0.001$). Notably, there were no hospital deaths. *Conclusions:* The development of double-chambered right ventricle in association with ventricular septal defect results in an increased pressure gradient within the right ventricle. The defect needs correction in a timely manner. In our experience, the surgical correction of double-chambered right ventricle is safe and shows excellent early and mid-term results.

Double-chambered right ventricle is a rare and progressive form of CHD which occurs due to muscle hypertrophy of the right ventricle.[1] Double-chambered right ventricle is further categorised as: Type 1-obstruction due to anomalous muscle bundle crossing right ventricle and Type 2-hypertrophy of parietal and septal muscle.[2] Clinical evidence reports that double-chambered right ventricle is highly associated with ventricular septal defect in 90% of the cases. Other associated anomalies include atrial septal defect, transposition of great vessels, tetralogy of Fallot, aortic regurgitation, Ebstein malformation, and ruptured sinus of valsalva aneurysm.[3] Double-chambered right ventricle is commonly reported, diagnosed, and treated during childhood and adolescence.[4] In an echocardiographic examination, a subcostal short-axis view is used in infants, and a parasternal short-axis view is used in adults to diagnose the development of double-chambered right ventricle.[5] However, surgical interventions such as transatrial, trans-ventricular, and corrective procedures treat the defect. Moreover, the transatrial approach is preferred to prevent postoperative arrhythmia.[6,7] Since 1960s, double-chambered right ventricle has been reported in clinical series. These reports suggest that the long-term results of surgical treatment are good with very less reintervention.[8,9] Based on this background, the present study aimed to review early and midterm outcomes of primary repair after double-chambered right ventricle in 64 patients over a period of 8 years.

Patients and methods

Patients

A total of 64 patients were diagnosed with double-chambered right ventricle and underwent surgical repair from January 2014 to June 2021in the departments of Cardiology and Cardiovascular and Thoracic Surgery of Postgraduate Institute of Medical Education and Research (PGIMER), Chandigarh, India. Written informed consent was obtained from all the subjects before their enrollment in the study.

Inclusion and exclusion criteria

The patients undergoing intra-cardiac repair for double-chambered right ventricle were included in the study. Patients who had double-chambered right ventricle with aortic override, pulmonary valvular, and main or branch pulmonary artery stenosis were excluded from the study.

Operative procedure

All the recruited patients underwent surgical correction for double-chambered right ventricle through a median sternotomy. For myocardial protection, 30 ml/kg of Delnido cardioplegia followed by 20 ml/kg was given to the patient every 60 minutes. A rectangular patch of the pericardium was harvested and treated with 0.6% glutaraldehyde for 6 minutes. Muscle band was excised through the right atrium in all the patients. Pulmonary valvotomy, patch augmentation of MPA, or RVOT were not required in any patient. The ventricular septal defect was closed with a poly-tetra-fluoro-ethylene/pericardial patch.

Data collection

This is a retrospective observational study. The data of the recruited patients included pre-operative (age, gender, co-existing anomalies such as ventricular septal defect, type of ventricular septal defect, and pressure gradient), intraoperative (aortic cross-clamp time, cardiopulmonary pass time, and hours of inotrope), and post-operative parameters (ventilation time, Vasoactive Inotropic Score, pressure gradient through trans thoracic echocardiography, ICU stay, and hospital stay). The data were collected by reviewing the clinical records, echocardiograms, and cardiac catheterisation studies of the patients. All the patients were subjected to follow-up and their clinical information was recorded. All patients were followed up once biweekly for 1 month initially, then once monthly for 3 months, once three monthly for 1 year, and thereafter. Echocardiography was done once in a year unless any complications. Data were collected from the electronic institutional database.

Statistical analysis

Data were expressed as mean with standard deviation. A 95% confidence interval was calculated for all the parameters. Normality was assessed using the KS test. Based on normality, a non-parametric approach was used for statistical analysis. The pre-operative and post-operative pressure gradients were compared and analysed using the Wilcoxon test. The statistical analysis was performed using GraphPad Prism v.8. The p-value of 0.05 was considered significant.

Results

Baseline characteristics and pre-operative outcomes

The baseline characteristics and pre-operative outcomes are summarized in Table 1. The mean age of the patients recruited was 13.42 ± 12.31 years. Of 64 patients, 36 (56.3%) were males and 28 (43.7%) were females. Echocardiography diagnosis revealed an associated co-anomaly i.e., ventricular septal defect in all the recruited subjects; 48 (75%) patients of sub-arterial type with a mean size of 10.52 ± 2.36 mm, 15 (23.4%) of perimembranous type with a mean size of 4.6 ± 1.88 mm and one (1.6%) patient of mid muscular type. The mean pre-operative pressure gradient was found to be 62.33 ± 5.52 mm of Hg. On intraoperative trans-oesophageal echocardiography, additional mid-muscular VSD was found in one patient and closed with direct suture closure. A Cath study or MRI was not done on any of the patient in this cohort.

Table 1. Baseline characteristics and pre-operative outcomes.

Parameters n (%)	Number of patients (n = 64)	95% CI
Age, years (Mean SD)	13.42 ± 12.31	10.35–16.50
• <1 year	02 (3.1)	
• 1–12 years	36 (56.2)	
• >12 years	26 (40.6)	
Sex		–
• Male	36 (56.3)	
• Female	28 (43.7)	
Preoperative medication		–
• Furosemide	64 (100)	
• Enalapril	14 (21.8)	
Preoperative NYHA status		–
• NYHA I	00 (0)	
• NYHA II	46 (71.9)	
• NYHA III	18 (28.1)	
• NYHA IV	00 (0)	
Preoperative ECG		–
• Right atrial enlarged	49 (76.5)	
• Bi-ventricular hypertrophy	09 (14.0)	
• Right ventricular hypertrophy	54 (84.3)	
• RBBB	28 (43.7)	
Preoperative CXR		–
• Right atrial enlargement	39 (60.9)	
• Left atrial enlargement	31 (48.4)	
• Right sided aortic arch	6 (9.3)	
Preoperative transthoracic echocardiography		–
• PFO/OS-ASD	19 (29.6)	
• PDA	11 (17.1)	
Type of VSD		
• Sub-arterial type	48 (75)	–
- Size, mm (Mean ± SD)	10.52 ± 2.36	9.83–11.21
• Perimembranous type	15 (23.4)	–
- Size, mm (Mean ± SD)	4.6 ± 1.88	3.56–5.64
• Mid-muscular, n (%)	01 (1.6)	–
Pressure gradient, mmHg (Mean ± SD) (Preop Transthoracic Echo)	62.33 ± 5.52	60.95–63.71

% = percentage, CI = confidence interval, CXR = cross-clamp rate, NYHA = New York heart association, n = number of patients, PFO = patent foramen ovale, SD = standard deviation, VSD = ventricular septal defect.

Intraoperative parameters

The mean times of aortic cross-clamp and cardiopulmonary bypass were found to be 81.58 ± 15.71 minutes and 106.58 ± 18.37 minutes, respectively. Further, the mean hours of inotrope were 37 ± 9.82. Mid-cavity obstruction was present in all patients and additional infundibular obstruction was present in 39 (60.9%) patients. Repeat muscle bundle resection was done in 5 (7.8%) patients due to increased right ventricular pressure and left ventricular pressure ratio. During repeat resection, trans right atrial approach was used.

https://doi.org/10.1017/S1047951123001531 Published online by Cambridge University Press

Table 2. Post-operative outcomes.

Parameters (Mean ± SD)	Number of patients (n = 64)	95% CI
Vasoactive-Inotropic Score	12.55 ± 3.33	11.72–13.38
Hours of ventilation	18.81 ± 4.61	17.66–19.96
ICU stay, days	5.94 ± 1.51	5.56–6.32
Hospital stay, days	9.11 ± 1.91	8.63–9.59
Postoperative pressure gradient, mmHg (Predischarge transthoracic echocardiography)	15.73 ± 2.94	15–16.46
Post-operative events, n (%)		–
• Low cardiac output syndrome	07 (10.9)	
• Junctional ectopic tachycardia	11 (17.1)	
• Reintubation	03 (4.6)	
• AKI	02 (3.1)	
Follow-up, months	46.73 ± 27.37	39.89–53.57
New York Heart Association (NYHA) status, n (%) (During last follow-up)		–
• NYHA I	24 (37.5)	
• NYHA II	39 (60.9)	
• NYHA III	01 (1.6)	

% = percentage; CI = Confidence interval; n = number of patients; SD = standard deviation.

Post-operative outcomes

All the surgical interventions were successful and there were no hospital or late deaths. The mean ventilation time post-operatively was 12.55 ± 3.33 hours. 49 (76.5%) patients required blood transfusion during the intraoperative and post-operative periods. Further, the mean ICU and hospital stays were 5.94 ± 1.51 days and 9.11 ± 1.91 days, respectively. After surgery, one patient had a complete heart block and an epicardial pacemaker was inserted during their hospital stay. The mean Vasoactive Inotropic Score of the patients was found to be 12.55 ± 3.33. The patients were subjected to follow-up for a period of 6–96 months and the mean follow-up period was 46.73 ± 27.37 months. Notably, no patient was lost to follow-up.

Further, the post-operative pressure gradient was 15.73 ± 2.94 mmHg. A significant decrease in the mean pressure gradient from 62.33 ± 5.52 mmHg pre-operatively to 15.73 ± 2.94 mmHg post-operatively was observed ($p < 0.001$), suggesting a favourable outcome of the surgical intervention.

Lastly, of 64 patients, only one (1.6%) patient had New York Heart Association (NYHA) III. Among others, 24 (37.5%) patients had NYHA I, and 39 (60.9%) had NYHA II. The post-operative outcomes are detailed in Table 2. During the follow-up, a tiny residual ventricular septal defect was observed in one patient. The mean gradient during the last follow-up was 14.18 ± 2.64 mmHg.

Discussion

Double-chambered right ventricle is a rare congenital anomaly that occurs due to muscle hypertrophy in the right ventricle. The most documented and acquired phenomenon of double-chambered right ventricle is an obstruction in the right ventricle due to the presence of AMB. Folger proposed a simple classification of double-chambered right ventricle describing that AMB divides the right ventricle into a high-pressure inflow chamber and a low-pressure outflow chamber.[10] Double-chambered right ventricle develops in 0.5–2% of all cases with CHD. Due to its rare occurrence, only 64 patients in a tenure of 8 years could be enrolled in the study. Case reports and case series in the literature reported that most of the cases are diagnosed in children and adolescents and few reports in adults. In a study, McElhinney et al. reported three adults with double-chambered right ventricle aged 38–63 years old.[11] Further, the prevalence of double-chambered right ventricle is more in males than females, as per the male-to-female predilection ratio.[12] In our study, there were 36 (56.3%) males and 28 (43.7%) females.

Double-chambered right ventricle is rarely observed as an isolated anomaly. It is most commonly associated with ventricular septal defect. The other co-anomalies include stenosis, tetralogy of the Fallot, double outlet right ventricle, and Ebstein anomaly.[13] In our study, all the patients with double-chambered right ventricle had an associative ventricular septal defect. Clinical evidence in the literature reported that 90% of cases are associated with the perimembranous type of ventricular septal defect.[14] Contrary to these reports, 48/64 (75%) patients represented sub-arterial type and 15 (23.4%) patients had perimembranous type of ventricular septal defect in our study. The isolated double-chambered right ventricle is asymptomatic; however, patients may experience symptoms in double-chambered right ventricle associated with ventricular septal defect.[15]

In a routine echocardiographic examination, the right ventricular outflow tract is not assessed due to difficulty in obtaining the image at the proximity of the outflow tract to a transducer. However, in the echocardiographic examination owing to double-chambered right ventricle, the images were obtained for the subcostal short-axis view in infants and the parasternal short-axis view in adults. The trans-thoracic and trans-oesophageal echocardiographic examinations are of diagnostic importance that help in reducing misdiagnosis of double-chambered right ventricle in patients.[5] Lastly, catheterisation and cardiac MRI are adjuncts to evaluate pressure gradients, right ventricular volume, and function consecutively.[3] This technique has an excellent spatial resolution. The presence of AMB causes a pressure gradient between the inflow and outflow portions of the right ventricle of the patients. The increase in pressure gradient acts as an initial stimulus for hypertrophy, resulting in the development of double-chambered right ventricle in human subjects.[14] The mean pre-operative pressure gradient was found to be 62.33 ± 5.52 mmHg, which is much higher than the normal range of ~ 20-30 mmHg. The pressure gradient tends to progress with age. [4,15] Therefore, early surgical intervention for double-chambered right ventricle correction and repair is highly recommended in these cases.

The surgical correction of double-chambered right ventricle involves surgical resection of the obstruction. Patients with elevated pressure gradients of more than 40 mmHg are suggested to undergo surgical intervention. However, the clinician or surgeon may elect to perform the surgery based on the condition of the patient i.e., symptomatic and high-pressure gradient.[16] In a few cases, beta-blockers have been used to improve the symptoms and can be considered helpful as pr-surgical adjuncts.[7] There are two main approaches i.e., transatrial and transventricular for the surgical correction of double-chambered right ventricle.[5] All the surgical interventions were successful as there were no hospital or late deaths. The mean follow-up period was 46.73 ± 27.37 months. Notably, no patient was lost during follow-up. During their follow-up, a significant decrease in the mean pressure gradient from 62.33 ± 5.52 mmHg pre-operatively to 15.73 ± 2.94 mmHg post-

https://doi.org/10.1017/S1047951123001531 Published online by Cambridge University Press

operatively was observed ($p < 0.0001$). Our findings are in accordance with the study carried out by Kahr et al (2014) that revealed positive outcomes after surgery with minimal mortality. These post-operative outcomes favour surgical intervention in patients with double-chambered right ventricle.[8]

Conclusion

Double-chambered right ventricle is a congenital heart anomaly that occurs mainly in childhood and rarely in adulthood. Diagnostic techniques such as echocardiography, catheterisation, and cardiac MRI are very effective in ensuring the development of double-chambered right ventricle. Early diagnosis leads to an early operational procedure to rectify the defect in an appropriate time frame. The surgical interventions result in excellent early and midterm outcomes.[17]

Acknowledgement. We are grateful to Dr. Rajni Bala for the statistical analysis.

Competing interests. None.

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https://doi.org/10.1017/S1047951123001531 Published online by Cambridge University Press