Cardiology in the Young

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Early and late outcomes of surgical repair of double-chambered right ventricle: a single-centre experience*

Murat Surucu[1], İlkay Erdoğan[2], Birgül Varan[2], Murat Özkan[3], N. Kürşad Tokel[2] and Sait Aşlamacı[3]

[1]Department of Pediatric Cardiology, University of Health Sciences, Diyarbakır Gazi Yasargil Training and Research Hospital, Diyarbakır, Turkey; [2]Department of Pediatric Cardiology, Faculty of Medicine, University of Başkent, Ankara, Turkey and [3]Department of Cardiovascular Surgery, Faculty of Medicine, Başkent University, Ankara, Turkey

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Author for correspondence:
M. Surucu, Department of Pediatric Cardiology, University of Health Sciences, Diyarbakır Gazi Yasargil Training and Research Hospital, Diyarbakır 21003, Turkey. Tel: +90 507 055 80 12. E-mail: pdr.murats@gmail.com

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Abstract

Objective: Double-chambered right ventricle is characterised by division of the outlet portion of the right ventricle by hypertrophy of the septoparietal trabeculations into two parts. We aim to report our experiences regarding the presenting symptoms of double-chambered right ventricle, long-term prognosis, including the recurrence rate and incidence of arrhythmias after surgery. *Methods:* We retrospectively investigated 89 consecutive patients who were diagnosed to have double-chambered right ventricle and underwent a surgical intervention from 1995 to 2016. The data obtained by echocardiography, cardiac catheterisation, and surgical findings as well as post-operative follow-up, surgical approaches, post-operative morbidity, mortality, and cardiac events were evaluated. *Results:* Median age at the time of diagnosis was 2 months and mean age at the time of operation was 5.3 years. Concomitant cardiac anomalies were as follows: perimembranous ventricular septal defect (78 patients), atrial septal defect (9 patients), discrete subaortic membrane (32 patients), right aortic arch (3 patients), aortic valve prolapse and/or mild aortic regurgitation (14 patients), and left superior caval vein (2 patients). The mean follow-up period was 4.86 ± 4.6 years. In these patients, mean systolic pressure gradient in the right ventricle by echocardiography before, immediately, and long-term after surgical intervention was 66.3, 11.8, and 10.4 mmHg, respectively. There were no deaths during the long-term follow-up period. Surgical reinterventions were performed for residual ventricular septal defect (2), residual pulmonary stenosis (1), and severe tricuspid insufficiency (1). *Conclusion:* The surgical outcomes and prognosis of double-chambered right ventricle are favourable, recurrence and fatal arrhythmias are unlikely in long-term follow-up.

Double-chambered right ventricle is a rare CHD, characterised by division of the right ventricle by hypertrophy of the septoparietal trabeculations into two parts, the proximal chamber with higher pressure and the distal chamber with low pressure.[1] Depending on the high right ventricular pressure, chest pain and heart failure symptoms may be seen, but most patients are asymptomatic. In addition to being asymptomatic, progressive course of the disease may cause delays in the diagnosis. Approximately 80–90% of the patients have associated ventricular septal defect. Double-chambered right ventricle diagnosis can be made at the same time as ventricular septal defect diagnosis, and some patients may progressively develop hypertrophy in muscle bands long-term follow-up after surgery with ventricular septal defect.[2,3] Although surgery is the only treatment option, there are some difficulties in deciding the timing. Surgery is recommended, even if in asymptomatic patients in the early stages, with the aim of preventing long-term exposure of the right ventricle to high pressure. Long-term follow-up results are important to evaluate the risk of recurrence after surgery and post-operative clinical course.

We aim to report our experience regarding the clinical presentation of double-chambered right ventricle, long-term prognosis, including the recurrence rate and incidence of arrhythmias after surgery.

Materials and methods

We retrospectively investigated 89 consecutive patients who were diagnosed to have double-chambered right ventricle and underwent a surgical intervention from 1995 to 2016. Clinical and operative data were extracted from medical records. We investigated the following clinical characteristics before and after surgery: gender; age at the time of surgery; follow-up period; right ventricle pressures and gradients measured by transthoracic echocardiography and cardiac catheterisation; and co-existing conditions, such as ventricular septal defect, atrial septal defect. The surgical approaches, post-operative mortality rate, and incidence of cardiac events were also evaluated.

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Table 1. Demographic characteristics of patients and initial diagnosis

Patients (n)	89 (30 Females, 59 Males)
Median age at the time of diagnosis	2 months (1 month to 30 years)
Mean age at the time of operation	5.3 ± 4.89 years (5 months to 30 years)
Initial diagnosis	Ventricular septal defect – 47.2%
	Ventricular septal defect + pulmonary stenosis – 20.2%
	Tetralogy of Fallot – 3.4%
	Pulmonary stenosis – 6.7%
	Ventricular septal defect + discrete subaortic membrane – 6.7%

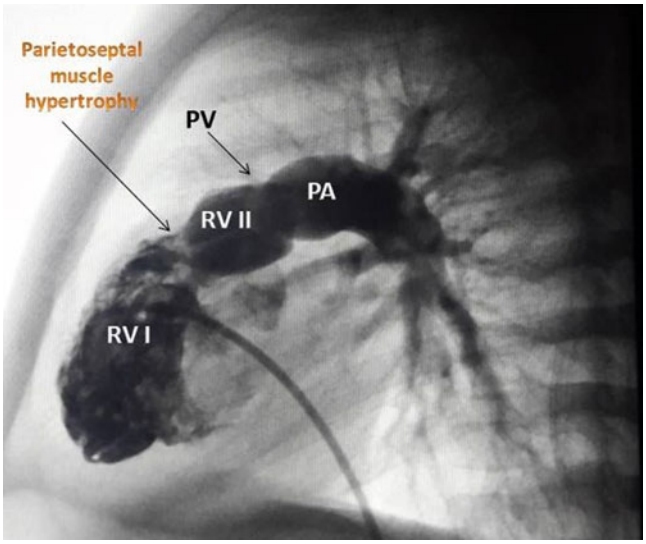

Figure 1. Angiography in lateral view shows the two chambers within the right ventricle divided by hypertrophied septoparietal musculature. PA = pulmonary artery; PV = pulmonary valve; RV I = proximal right ventricular chamber; RV II = distal right ventricular chamber.

Transthoracic echocardiography was performed before and immediately after the surgical intervention and during long-term follow-up. Cardiac catheterisation was performed in 63 patients (70.7%) to show the location of the stenosis in the right ventricle and to record the pressure (Fig 1).

Diagnostic criteria for double-chambered right ventricle were (1) echocardiographic or angiographic evidence of a mid-ventricular obstruction (a systolic pressure gradient between the right ventricle proximal chamber (inflow) and outflow tracts) and (2) absence of infundibular hypoplasia.

Statistical analyses were performed using SPSS for Windows 22.0 (SPSS, Chicago, IL, United States of America). Data are expressed as mean ± standard deviation or median (range) values.

This study was approved by Başkent University Institutional Review Board (project no: 94603339-604.01.02/22872).

Results

A total of 89 patients (59 males and 30 females) were diagnosed with double-chambered right ventricle and underwent a surgical intervention from 1995 to 2016. Median age at the time of diagnosis was 2 months and mean age at the time of operation was 5.3 years (Table 1). The presenting symptoms were murmur on physical examination in 80 patients, cyanosis in 1, chest pain in 2, fatigue in 1, syncope in 1, and shortness of breath in 1. Concomitant cardiac anomalies were as follows: perimembranous ventricular septal defect (78 patients), muscular ventricular septal defect (2 patients), atrial septal defect (9 patients), discrete subaortic membrane (32 patients), right aortic arch (3 patients), aortic valve prolapse and/or mild aortic regurgitation (14 patients), left superior caval vein (2 patients), ductus arteriosus (1 patient), and peripheral pulmonary stenosis (1 patient). The initial diagnosis of patients before being referred to our centre was not double-chambered right ventricle in 84.2%. Double-chambered right ventricle progressively developed in three patients who were operated with the diagnosis of atrial septal defect + pulmonary stenosis, ventricular septal defect, and ventricular septal defect + discrete subaortic membrane, respectively.

The mean follow-up period was 4.86 ± 4.6 years (1 month to 17 years). According to the patients' clinical and laboratory findings in the follow-up period, echocardiography and electrocardiography were performed in the 1st, 3rd, and 6th month after the operation and then once a year.

All patients were operated with cardiopulmonary bypass, and cold crystalloid cardioplegia was used to establish diastolic cardiac arrest. The right ventricular hypertrophic bands were resected via tricuspid valve in 29 patients, depending on the age of the patient and the extent of obliteration of the right ventricle cavity. In the rest of the patients, a right ventricular incision with resection via the ventriculotomy was necessary. In 11 patients, the right ventriculotomy was closed primarily. A limited bovine pericardial or autologous pericardial infundibular patch was used to close the right ventriculotomy in order to break a circumferentially contracting outflow tract ring for the rest of the patients. The main concern in incorporating an infundibular patch has been to avoid recurrent outflow tract stenoses. A transannular patch was necessary in only four patients. Thirty-two patients were noted to have discrete subaortic membrane or ridge; of these, 22 were considered to be significant and resected during the surgery.

In these patients, the systolic pressure gradient in the right ventricle by echocardiography before, immediately, and long-term after surgical intervention was 66.3 ± 28 (26–168), 11.8 ± 7.9 (2.8–44), and 10.4 ± 6.9 (3–40) mmHg, respectively. The post-operative echocardiography revealed residual ventricular septal defect in 33 patients which were noted as insignificant, residual pulmonary stenosis in 2 patients, and coronary arteriovenous fistula in 15 patients in the right ventricular outflow tract. Mean post-operative ICU stay was 3.1 ± 2 (1–15) days and mean hospital stay was 7.8 ± 5 (3–40) days. Re-exploration for bleeding was required in one patient and one patient required plication of the diaphragm due to phrenic nerve injury. Four patients (4.5%) required surgical reinterventions.

Within the early post-operative period, six patients required temporary pacemakers for atrioventricular block, and three of them had a permanent epicardial pacemaker implantation. The first patient with a permanent pacemaker had a ventricular septal defect closure and developed double-chambered right ventricle during the follow-up. The second patient underwent ventricular septal defect closure, discrete subaortic membrane resection, hypertrophic band resection at age of 8 years. The third patient underwent ventricular septal defect closure and right pulmonary artery reconstruction as well as hypertrophic band resection.

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Table 2. Prognosis and cardiac events during follow-up period

Long-term prognosis	N (%)
All cause death	–
Mean post-operative ICU stay (day)	3.1 ± 2.3
Mean hospital stay (day)	7.8 ± 5.2
Post-operative echocardiography	
Residual VSD	33 (37%)
Pulmonary stenosis	2 (2.2%)
Coronary arteriovenous fistula	15 (16.8%)
Surgical reintervention	
Residual VSD	2 (2.2%)
Residual pulmonary stenosis	1 (1.1%)
Severe tricuspid insufficiency	1 (1.1%)
Arrhythmias	
Complete atrioventricular block	3 (3.3%)
Premature ventricular contraction	3 (3.3%)
Supraventricular tachycardia	1 (1.1%)

VSD = ventricular septal defect.

Throughout the follow-up period, a 24-hour rhythm Holter were performed in 18 patients with palpitations in the outpatient clinic follow-up or with arrhythmia on electrocardiography. A 24-hour rhythm Holter study on long-term follow-up revealed frequent ventricular extrasystoles in three patients; however, only one of them required medication. There was no death during the follow-up period in this patient cohort (Table 2).

Discussion

Double-chambered right ventricle is a rare anomaly in all CHDs with a rate of 0.5–2%.[4] Double-chambered right ventricle is considered to be a progressive CHD that develops due to hypertrophy of the abnormal muscle band in the right ventricle. Since there is not enough data, it is difficult to indicate the presence of a genetic susceptibility or acquisition of the disease. However, in similar case studies, it has been reported that patients undergoing surgery of ventricular septal defect may develop double-chambered right ventricle and acquire the disease.[5,6] In our study, three patients had previously undergone operation, including atrial septal defect + pulmonary stenosis, ventricular septal defect, and ventricular septal defect + discrete subaortic membrane, respectively, and progressively developed double-chambered right ventricle in follow-up.

Double-chambered right ventricle includes three main forms including isolated, with an associated ventricular septal defect, and with an associated subaortic stenosis.[7-9] A ventricular septal defect is present in greater than 50–80% of cases and usually perimembranous. Most of the times, the ventricular septal defect is small and tends to a spontaneous closure. When septal defect is due to malalignment, there seems to be a higher tendency towards left ventricular outflow tract obstruction, which is usually due to a discrete subaortic stenosis.[10] In this study, 87.6% of patients had perimembranous ventricular septal defect, 2.2% of muscular ventricular septal defect. Discrete subaortic membrane resection was performed in 22 out of 32 patients with subaortic membrane. Nine patients with ventricular septal defect showed spontaneous closure in the pre-operative period. One of them was muscular type, while others were perimembranous type.

Although double-chambered right ventricle is well recognised in childhood, it may be misdiagnosed in adult patients. Transthoracic and/or transoesophageal echocardiography are the methods of choice for the diagnosis of double-chambered right ventricle. Due to limitations of echocardiography in adult patients, this entity may be missed, particularly if it presents concomitant with other congenital defects. Therefore, additional imaging methods such as MRI or cardiac catheterisation are required for a definitive diagnosis.

When a right mid-ventricular obstruction is detected, a warning sign should switch on in the operator's mind, and a careful evaluation of the membranous ventricular septum and the subaortic region should be carried out. In this study, a diagnosis of double-chambered right ventricle was made at the end of angiography in four patients followed by a diagnosis of ventricular septal defect.

In patients diagnosed with double-chambered right ventricle, surgical treatment should be planned when the peak pressure difference with cardiac catheterisation between low and high placement chambers reaches 40 mmHg, or if the pressure difference is less than 40 mmHg in the presence of ventricular septal defect with heart failure symptoms. The period for surgical repair usually depends on the associated cardiac anomalies. In the absence of a significant coexisting defect, observation is possible as long as the intracavitary systolic gradient is not higher than 40 mmHg and the obstruction is not progressive. Surgical correction of double-chambered right ventricle results in excellent functional and haemodynamic long-term results, with a complete relief of the right ventricular obstruction. In this study, 63 patients underwent pre-operative cardiac catheterisation, and 25 patients with a pressure difference of less than 40 mmHg between the chambers had a ventricular septal defect and septal defect closure and hypertrophic band resection due to the presence of heart failure symptoms. In 38 patients, the pressure difference was higher than 40 mmHg.

Various surgical techniques have been described for resection of the obstructing anomalous muscle bundles from the right ventricle cavity, including transventricular, transatrial, or combined approaches.[9] In our study, the transventricular approach was used in 60 patients (67.5%) and the transatrial in 29 patients (32.5%). The transatrial approach is safer in terms of right ventricular dysfunction and arrhythmia risk in long-term follow-up compared to the transventricular approach. However, right ventriculotomy approach may have an advantage, especially in severe right ventricle obstruction. The right ventriculotomy approach permits adequate relief of right ventricular cavity obstruction, allowing visualisation of the ventricular septal defect margin, which is sometimes difficult to identify through a transatrial approach in the presence of heavy obstructing right ventricular muscle bundles.

The incidence of aortic valve insufficiency has been reported in up to 40% in adult double-chambered right ventricle patients and in 5–20% in paediatric patients.[10] Aortic valve prolapse and valve insufficiency are documented more frequently in double-chambered right ventricle patients than patients with ventricular septal defect only.[11] In our series, aortic valve prolapse and/or mild aortic regurgitation were detected in 14 patients (aortic valve prolapse in 6 patients, aortic valve prolapse + mild aortic valve insufficiency in 2 patients, and only mild aortic valve insufficiency in 6 patients). The mean age of the patients was 5.96 ± 5.44 years. Due to the mild insufficiency, no surgical intervention was performed for aortic valve.

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Surgical correction and long-term follow-up of double-chambered right ventricle result in excellence. In this series, there were no deaths during the long-term follow-up period. Surgical reinterventions were performed for residual ventricular septal defect with infective endocarditis (two patients, 3 months later), residual pulmonary stenosis (one patient, 6 years later), and severe tricuspid insufficiency (one patient, 3 years later). Right ventricular dysfunction and lethal arrhythmias were rarely reported in the follow-up of double-chambered right ventricle patients with frequent premature ventricular contractions in three patients in our series. In addition, three patients had permanent pacemaker implantation following a complete atrioventricular block development.

Conclusions

It is important to be aware of the presence of ventricular septal defect and discrete subaortic membrane when evaluating patients who will undergo double-chambered right ventricle surgery since it can increase early post-operative morbidity and may require a second surgical intervention if it is not diagnosed in the pre-operative period.

The surgical outcomes and post-operative prognosis of double-chambered right ventricle are favourable, and neither recurrence of double-chambered right ventricle nor fatal arrhythmias develop during the long-term follow-up period.

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Conflicts of Interest. None.

References

1. Loukas M, Housman B, Blaak C, Kralovic S, Tubbs RS, Anderson RH. Double-chambered right ventricle: a review. Cardiovasc Pathol 2013; 22(6): 417–423. doi: 10.1016/j.carpath.2013.03.004.
2. Alva C, Ho SY, Lincoln CR, Rigby ML, Wright A, Anderson RH. The nature of the obstructive muscular bundles in double-chambered right ventricle. J Thorac Cardiovasc Surg 1999; 117 (6): 1180–1189.
3. Pongiglione G, Freedom RM, Cook D, Rowe RD. Mechanism of acquired right ventricular outflow tract obstruction in patients with ventricular septal defect: an angiocardiographic study. Am J Cardiol 1982; 50(4): 776–80.
4. Hoffman P, Wójcik AW, Rózański J, et al. The role of echocardiography in diagnosing double chambered right ventricle in adults. Heart 2004; 90: 789–793.
5. Işık O, Akyüz M, Ayık MF, Atay Y. Late development of double-chambered right ventricle after repair of a ventricular septal defect. Turk Kardiyol Dern Ars 2015; 43(7): 640–643. doi: 10.5543/tkda.2015.50870.
6. Sherif M, David JP, Nanette A, et al. Double chambered right ventricle with ventricular septal defect in adults: case series and review of the literature. J Cardiovasc Ultrasound 2015; 23(1): 48–51. doi: 10.4250/jcu.2015.23.1.48.
7. Freedom RM, Yoo SJ, Mikailian H, Williams WG. The natural and modified history of congenital heart disease. New York, USA: Wiley-Blackwell, 2004.
8. Barik R. Role of echocardiograghy in treating a case of double chamber right ventricle with delayed presentation. J Cardiovasc Echography 2017; 27: 10–3.
9. Penkoske PA, Duncan N, Collins-Nakai RL. Surgical repair of double-chambered right ventricle with or without ventriculotomy. J Thorac Cardiovasc Surg 1987; 93: 385–393.
10. Hachiro Y, Takagi N, Koyanagi T, Morikawa M, Abe T. Repair of double-chambered right ventricle: surgical results and long-term follow-up. Ann Thorac Surg 2001; 72(5): 1520–1522.
11. Wang JK, Wu MH, Chang CI, et al. Malalignment-type ventricular septal defect in double-chambered right ventricle. Am J Cardiol 1996; 77(10): 839–842.