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SURGICAL EXPERIENCE WITH DOUBLE CHAMBER RIGHT VENTRICLE PRESENTED WITH RIGHT VENTRICLE OUTFLOW TRACT OBSTRUCTION

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Contribution

TW conceived, designed the research methodology, prepared the manuscript and is accountable for the originality of the research work. ZA did data collection, helped in writing the manuscript and reviewed the manuscript. Both authors contributed significantly to the submitted manuscript.

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ABSTRACT

Objective: To present our surgical experience with double chamber right ventricle pathology.

Methodology: This cross sectional study included retrospective analysis of cardiac surgery database at Chaudhry Pervaiz Elahi Institute of Cardiology Multan from May 2010 to April 2016. This study included surgical repair of patients having either right ventricle outflow tract or mid cavity obstruction due to muscle bundles leading to DCRV. Cases of Pulmonary Stenosis, Tetralogy of Fallot, Pulmonary Atresia and Double Outlet Right Ventricle causing RVOT obstruction were excluded from study.Data was analyzed using MS Excel. Mean and Frequency were calculated for quantitative and qualitative variables respectively.

Results: Out of twenty-five patients, 36% patients had moderator band and 64% patients had anomalous muscle bands responsible for DCRV and main presentation was right ventricle outflow tract obstruction (RVOT PG = 85.64 $\pm$48.91mmHg). Moreover, 12% of patients had Atrial Septal Defect and 52% had Ventricular Septal Defect as associated cardiac defects. Surgical repair results were excellent with no operative mortality.

Conclusion: The main clinical presentation in our Double Chamber Right Ventricle patients was right ventricle outflow tract obstruction either caused by a moderator band or anomalous muscle bundles. Surgical repair of DCRV and associate cardiac defects carry excellent results with low morbidity.

Key Words: Double chamber right ventricle, Right ventricle outflow tract obstruction, Moderator band, Anomalous muscle bundle, Ventricle septal defect

INTRODUCTION

Double Chamber Right Ventricle (DCRV) is a rare clinical entity encountered in pediatric cardiac surgery.[1] Clinical presence is often missed in presence of associated cardiac defect like VSD in majority of case and inability to view standard trans-thoracic echocardiographic views i.e. apical four chamber view.[2] Muscle bands causing DCRV need reoperation for persistent RV obstruction if not dealt at primary surgery.[3] Focused Echocardiographic view or cardiac MRI are warranted preoperatively for through search of this cardiac pathology to avoid less than complete correction of cardiac anomaly in suspected cases.[4]

DCRV is caused by RV muscle bands that divide the Right ventricle into two chambers with physiological separate hemodynamic parameters by creating pressure gradient. Usually proximal RV chamber has high pressures and hence thick walled as compared to distal chamber with low pressures and thin wall. Ventricle septal defect (VSD) is reported in up to 63-90% patients with DCRV.[5] Communication of VSD with high or low pressure RV chamber determine clinical picture. If Left Ventricle (LV) communicates with low-pressure chamber through VSD, then it behaves like simple VSD with Left to Right Shunt. While in case if it communicates with high pressure RV chamber, it appears like TOF having Right to Left Shunt. It is not uncommon to find that anomalous muscle bundle is also responsible for progressive obstruction to right ventricle outflow tract (RVOT).[6]Muscle bands responsible for DCRV are usually anomalous muscle band in RV cavity or misplaced moderator band.

Objective of our publication was to share our surgical experience with DCRV. We are presenting the frequency of associated cardiac anomalies, clinical presentation of DCRV and our surgical approach to the pathology.

METHODOLOGY

This was a cross sectional study conducted by retrospectively analyzing Cardiac Surgery Department Electronic Database to review operative finding of surgical procedure, clinical presentation and surgical outcome of interest. Data from May 2010 to April 2016 for patients operated for DCRV at CPE institute of Cardiology, Multan was analyzed. This analysis was reinforced by patient hospital paper data to recheck clinical information. Patients having either right ventricle outflow tract or mid cavity obstruction due to muscle bundles leading to DCRV were included in this study. Cases of pulmonary stenosis, Tetralogy of Fallot, pulmonary atresia and double outlet right ventricle causing RVOT obstruction were excluded. Age, gender, weight, height, NYHA class, associated cardiac anomalies on echocardiography along with pressure gradient across the RVOT of patient population were noted. Among the operative details; information regarding cardiac approach to muscle band resection, location of anomalous muscle band and associated surgical procedure were retrieved. All patients were operated on standard cardiopulmonary bypass with bicaval cannulation. Cold blood cardioplegia was used for arresting of heart. Moderate hypothermia was used for myocardial protection. DCRV pathology was approached through either right atrial (RA) approach or combine right atrial and pulmonary artery approach. Right ventriculotomy was avoided. Operative finding related to moderator band or anomalous muscle bundles causing DCRV and right ventricle outflow tract obstruction (RVOTO) were consistent with echocardiographic information. Adequacy of muscle resection and release of RVOTO was assessed per-operatively by passing appropriate size Hegar's dilator from RV to PA.

Postoperative echocardiography of every patient was done to check for presence of RVOT gradient, residual VSD and ventricular functions. Length of hospital stay and operative mortality were recorded from hospital record.

RESULTS

About 25 patients were operated for surgical repair of DCRV. Their baseline characteristics showed patient population had age of 15.04 $\pm$ 8.67 years. There were 19 (76%) males. Mean weight was 31.88 $\pm$ 13.40 kg, height 128.2$\pm$ 46.60 cm. About 18 patients belong to NYHA II while 7 were classified as having NYHA III as shown in Table 1. It is interesting to mention that around 50% of above mentioned patient population was primarily investigated for VSD or ASD work-up. ASD was present in 3 (12%) and 13 (52%) had VSD as associated cardiac defects. Among the patients who had VSD, all patients had peri-membraneous type VSD with 9 (36%) having right coronary cusp prolapsed leading to mild aortic valve regurgitation. Focus echocardiographic evaluation showed 9 patients

Table1: Preoperative Demographic Variables and Echocardiographic findings in Study Population (n=25)

Variable	Value
Age (Years)	15.04 ± 8.67
Weight (kg)	31.88 ± 13.40
Height (cm)	128.2± 46.60
GENDER	
Male	76.0% (n=19
Female	24.0% (n=6)
NYHA CLASS	
I	0.0%
II	72.0% (n=18)
III	28.0% (n=7)
IV	0.0%
Echocardiographic finding of muscle band	
Moderator Band	36.0% (n=9)
Anomalous Muscle Bundle	64.0% (n=16)
RVOTO	92% (n=23)
RVOT Pressure Gradient(mmHg)	85.64 ± 48.91
Associate cardiac defects	
ASD	12% (n=3)
VSD	52% (n=13)

NYHA=New York Heart Association, RVOTO=Right Ventricle Outflow Tract Obstruction, ASD=Atrial Septal Defect, VSD= Ventricular Septal Defect. ± indicates Standard deviation

(36%) had moderator band as cause of DCRV causing RVOT obstruction (RVOTO) while anomalous muscle band was involved in 16 cases (64%). RVOTO was present in 24 patients (92%) with mean pressure gradient of 85.64 ± 48.91mmHg. One patient primarily had severe right ventricle mid-cavity obstruction with RVOTO.

Postoperative echocardiography was done to reassess the adequacy of surgical repair. Postoperative echocardiography showed RVOT pressure gradient of 22.48± 11.88 mmHg. Length of hospital stay was 7.56± 1.04 days with 0% operative mortality (Table 2).

Table 2: Operative Approach, Length of Hospital Stay, Operative Mortality and Postoperative RVOT Pressure Gradient in Study Population (n=25)

Variable	Value
Surgical approach	
a. Right Atrial (RA)	64%(n=16)
b. Right Atrial + Pulmonary Artery (RA+PA)	36%(n=9)
Hospital stay(days)	7.56 ± 1.04
Operative mortality	0.0%
RVOT Pressure Gradient (mmHg)	22.48 ± 11.88

RVOT= Right Ventricle Outflow Tract,± indicates Standard deviation

DISCUSSION

Double chamber right ventricle is rare condition with progressive course and unknown natural history.[7] Associated cardiac anomalies i.e. VSD, TOF, ASD make situation more complex. Muscle bands usually responsible for creation of DCRV are either anomalous muscle band or displaced hypertrophied moderator bundle. In our population, presence of associated cardiac defect like VSD, ASD are similar as present in published data. But clinically remarkable presentation in our DCRV population was RVOTO in contrast to previously published data. Peri-membranous VSD was communicating low pressure chamber of DCRV and significant number of patient had prolapse of RCC resulting mild aortic valve regurgitation.[8,9] Resection of muscle bundles repaired the DCRV pathology. Aggressive resection is usually not advisable as it may lead to ventricle arrhythmia or late onset progressive RV dysfunction.[10] This situation become challenging in presence of TOF pathology. Inadequate muscle resection or failure to appreciate presence of DCRV will result in persistent high RV pressure or DCRV and may warrant surgical resection.[3]

Trans-atrial, trans-ventricle or combine trans-atrial & trans-ventricle approaches are utilize for correction of DCRV pathology.[11-14] In adult and grown up children with DCRV, trans-ventricle approach is preferred by some centers because of ease in identifying the muscle band and associated VSD.[15] Trans-atrial approach was used in majority of patients while in only few patients we used trans pulmonary approach. Similarly, Trans arterial approach was also preferred by Galal O et al.[16] In our population, relief of RVOTO and resection of muscle was satisfactory with improved postoperative RVOT pressure gradients. Right atrial + Pulmonary arteriotomy approach for DCRV repair along with associated cardiac defect carries very low operative mortality along with short hospital stay.

LIMITATIONS:

This study is based on single center experience with small number of patient. DCRV has frequency of less than 1% of all congenital cardiac defects. So, small study population is justifiable. Study is lacking cardiac catheterization data from proximal and distal chamber of DCRV. Actually, cardiac catheterization is not included in hospital work-up protocol for VSD, ASD or RVOTO unless advanced pulmonary hypertension is suspected or clinical picture of cardiac defect is confusing.In addition, Echocardiographic evaluation provided sufficient clinical information regarding muscle bands causing DCRV, ASD,VSD,ROVOTO.

CONCLUSION

In our DCRV population, main clinical presentation was right ventricle outflow tract obstruction that was most commonly caused either by a moderator band or anomalous muscle bundles. Surgical repair of DCRV and associate cardiac defects carries excellent surgical outcome.

REFERENCES

1. Kahr PC, Alonso-Gonzalez R, Kempny A, et al. Long-term natural history and postoperative outcome of double-chambered right ventricle--experience from two tertiary adult congenital heart centres and review of the literature. Int J Cardiol. 2014 Jul 1. 174 (3):662-8.
2. Hoffman P, Wojcik AW, Rozanski J, et al. The role of echocardiography in diagnosing double chambered right ventriclein adults. Heart. 2004 Jul. 90(7):789-93
3. Moran AM, Hornberger LK, Jonas RA, et al. Development of a double-chambered right ventricle after repair of tetralogyofFallot. J Am CollCardiol 1998;31:1127–33.
4. http://emedicine.medscape.com/article/889517-workup#c10
5. Alva C, o SY, Lincoln CR, et al. The nature of the obstructive muscular bundles in double-chambered right ventricle. J Thorac Cardiovasc Surg 1999;117:1180 –9.
6. 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 Oct. 50(4):776-80
7. Fellows KE, Martin EC, Rosenthal A. Angiocardiography of obstructing muscular bands of the right ventricle. AJR Am J Roentgenol 1977;128:249 –56.
8. Wong PC, Sanders SP, Jonas RA, et al. Pulmonary valve–moderator band distance and association with development of double-chambered right ventricle. Am J Cardiol 1991;68:1681–6.
9. Wang JK, Wu MH, Chang CI, et al. Malalignment-type ventricular septal defect in double-chambered right ventricle.Am J Cardiol 1996;77:839–42.
10. Harrison DA, Harris L, Siu SC, et al. Sustained ventricular tachycardia in adult patients late after repair of tetralogy of Fallot. J Am CollCardiol 1997 ; 30:1368 –73.
11. Hartmann AF, Goldring D, Ferguson TB, et al. The course of children with the two-chambered right ventricle. J Thorac Cardiovasc Surg 1970;60:72–83.
12. Li MD, Coles JC, McDonald AC. Anomalous muscle bundle of the right ventricle. Its recognition and surgical treatment. Br Heart J 1978;40:1040 –5.
13. Wallsh E, Weinstein G, Franzone AJ, et al. Anomalous right ventricular muscle bundle. Repair through the right atrium. J Thorac Cardiovasc Surg 1975;69:830–4.
14. Penkoske PA, Duncan N, Collins-Nakai RL.

Surgical repair of double-chambered right ventricle with or without ventriculotomy. J Thorac Cardiovasc Surg 1987;93:385–93.

15. Hachiro Y, Takagi N, Koyanagi T, et al. Repair of doublechambered right ventricle: surgical results and long-term follow-up. Ann ThoracSurg 200;72:1520 –2.

16. Galal O, Al-Halees Z, Solymar L, et al. Double-chambered right ventricle in 73 patients. Spectrum of the disease and surgical results of transatrial repair. Can J Cardiol 2000;16:167–744