**Introduction**

Double-Chambered Right Ventricle (DCRV) is a rare form of right-sided intraventricular obstruction. The obstructing muscle bundles are assumed to be congenital in nature, are commonly associated with perimembranous ventricular septal defects (VSD) and progress with time in most cases. Due to the association with VSDs, the majority of patients with DCRV are identified and treated surgically during childhood. However, DCRV may also be firstly diagnosed in adult patients during routine examinations or with onset of symptoms. With a small number of reports available for the adult population, open questions are remaining concerning the optimal treatment for these patients:

- Is right ventricular myectomy a safe and effective form of treatment?
- What is the long-term outcome after surgery?
- Should asymptomatic patients be treated surgically?

**Methods**

A retrospective review of medical records was carried out on all patients with the diagnosis of DCRV who presented to the outpatient clinics at the Royal Brompton Hospital, London, and the University Hospital Münster (January 2000 - March 2012).

**Findings**

In total, 53 Patients were identified in London and Münster (see below), with a median age of 39 years at the time of data analysis. The majority of patients had undergone surgical repair (68%). The diagnosis was established in all cases according to published criteria.

**Associated Lesions on Imaging**

Isolated DCRV is a rare finding (see below). In almost all cases, a VSD or history of VSD was present. In addition, 15 patients (28%) had right bundle branch block (RBBB) and 2 patients were in atrial fibrillation prior to surgery. 6 patients (11%) had Trisomy 21, there was one patient with CHARGE syndrome and Marfan syndrome, respectively.

**Surgical Indications**

All patients undergoing surgery had symptoms related to their right ventricular obstruction. Of the 17 patients that did not undergo surgical repair of their DCRV, 8 were asymptomatic, 1 patient refused surgery and 8 patients were in the process of evaluation for DCRV repair.

**Surgery**

Patients underwent surgery at a median age of 27 years (14 months to 67 years) after elective admission. Surgical access was achieved via right ventriculotomy in 35% of patients, by a combination of right atriotomy plus right ventriculotomy (26%) or pulmonary arteriotomy (22%), or by using a transatrial approach only (17%). In general, right ventriculotomies were avoided in surgeries after 2001. All concomitantly present defects were corrected at the same time.

**Immediate Post-Operative Course**

In general, patients withstood the surgical procedures very well. There were no hospital or late deaths in the study population. All patients were extubated in less than 48 hours. 2 patients required dual chamber-pacemaker insertion for complete heart-block.

**Follow-Up of Postoperative Patients**

Mean follow-up time after DCRV repair was 4 ± 9 years (1.5 months to 33 years). 31 patients (86%) remained free from any intra-ventricular obstruction during follow-up, 5 patients retained gradients < 30 mmHg, but no patient required re-operation. Symptoms improved in all operated patients immediately after surgery, particularly with regard to their subjective exercise capacity. In the long-term, 25 patients (69%) remained completely asymptomatic and enjoyed good quality of life. However, 8 patients remained in NYHA class II.

Findings on their latest TTE investigations, 10 ± 8 years postoperatively (6 months to 22 years) showed residual intraventricular gradients and other findings in a proportion of patients (see below). Most patients remained in sinus rhythm postoperatively, while postoperative RBBB was frequent (56%).

**Postoperative Findings on Imaging and ECG**

Follow-Up of Unoperated Patients

Unoperated Patients were followed-up for 8 ± 10 years. During this time, 8 patients remained fully asymptomatic despite significant intraventricular gradients (up to 100 mmHg). 2 patients presented with atrial fibrillation, but there was no evidence of ventricular arrhythmias.

**Surgical Myectomy** is a safe and effective method to decrease the level of right ventricular obstruction and symptoms in DCRV patients. No re-emergence of DCRV after surgical myectomy was found in our patient cohort.

Asymptomatic patients may be managed conservatively without significant complications. However, a proactive treatment approach appears to be warranted.

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No Disclosures.