

Surgical Treatment of Double Outlet Right Ventricle Complicated by Pulmonary Hypertension

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Abstract

Background: Double outlet right ventricle (DORV) is a group of complex congenital heart abnormalities. Preoperative pulmonary hypertension (PH) is considered an important risk factor for early death during the surgical treatment of DORV. The aim of this study was to report our experience on surgical treatment of DORV complicated by PH.

Methods: From June 2004 to November 2016, 61 patients (36 males and 25 females) aged 2 weeks to 26 years (median: 0.67 years and interquartile range: 0.42–1.67 years) with DORV (two great arteries overriding at least 50%) complicated by PH underwent surgical treatment in our center. All patients were categorized according to surgical age and lesion type, respectively. Pulmonary artery systolic pressure (PASP), pulmonary artery diastolic pressure (PADP), and mean pulmonary artery pressure (mPAP) were measured directly before cardiopulmonary bypass (CPB) was established and after CPB was removed. An intracardiac channel procedure was performed in 37 patients, arterial switch procedure in 19 patients, Rastelli procedure in three patient, Senning procedure in one patients, and Mustard procedure in one patient. The Student's *t*-test and Chi-squared test were performed to evaluate clinical outcomes of the surgical timing and operation choice.

Results: Fifty-five patients had uneventful recovery. PASP fell from 55.3 ± 11.2 mmHg to 34.7 ± 11.6 mmHg ($t = 14.05, P < 0.001$), PADP fell from 29.7 ± 12.5 mmHg to 18.6 ± 7.9 mmHg ($t = 7.39, P < 0.001$), and mPAP fell from 40.3 ± 10.6 mmHg to 25.7 ± 8.3 mmHg ($t = 11.85, P < 0.001$). Six (9.8%) patients died owing to complications including low cardiac output syndrome in two patients, respiratory failure in two, pulmonary hemorrhage in one, and sudden death in one patient. Pulmonary artery pressure (PAP) dropped significantly in infant and child patients. Mortality of both infants (13.9%) and adults (33.3%) was high.

Conclusions: PAP of patients with DORV complicated by PH can be expected to fall significantly after surgery. An arterial switch procedure can achieve excellent results in patients with transposition of the great arteries type. Higher incidence of complications may occur in patients with ventricular septal defect (VSD) type before 1 year of age. For those with remote VSD type, VSD enlargement and right ventricle outflow tract reconstruction are usually required with acceptable results. The degree of aortic overriding does not influence surgical outcome.

Key words: Congenital Heart Disease, Double Outlet Right Ventricle, Pulmonary Hypertension

INTRODUCTION

Double outlet right ventricle (DORV) is a group of complex congenital heart abnormalities, in which both the pulmonary artery and the aorta arise primarily from the right ventricle.^[1] These two great arteries may override the ventricular septum by more than 50%, 90%, or both arteries may arise fully from the right ventricle (200%).^[2,3] DORV is frequently associated with other intracardiac or extracardiac malformations. Four types of DORV (tetralogy of Fallot [TOF] type, transposition of the great arteries [TGAs] type, ventricular septal defect [VSD] type, and remote VSD type) have been classified, and different procedures described for the

different types of DORV, with varied rates of success and frequently high mortality.^[3-9]

These four types differ significantly in their surgical treatment, postoperative management, and prognosis. Except for TOF

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type, which usually has pulmonary hypotension (PH), the other three types frequently induce PH. Preoperative PH was concerned as an important risk factor for early deaths in the surgical treatment of DORV.^[8] To the best of our knowledge, large series of patients with DORV complicated by PH had been seldom reported in the literature. Therefore, in this study, by analyzing and summarizing the clinical data of surgical treatment of patients with DORV complicated by PH, the proper surgical timing, surgical technique choice, and surgical outcomes were explored.

METHODS

Patients

This retrospective study analyzed the clinical data of patients with DORV (two great arteries overriding at least 50%) complicated by PH (mean pulmonary artery pressure [mPAP] ≥ 25 mmHg [1 mmHg = 0.133 kPa]) receiving surgical treatment in the Department of Cardiology, the First Hospital of Tsinghua University, Beijing, China. Informed consent of surgical procedure was obtained from patients and their families before surgery. The study was approved by the Medical Ethical Committee of the First Hospital of Tsinghua University. From June 2004 to November 2016, a total of 61 patients (36 males and 25 females), aged 2 weeks to 26 years (median: 0.67 years and interquartile range [IQR]: 0.42–1.67 years) and with body weight 2.5–62.0 kg (median: 6.6 kg and IQR: 5.6–10.0 kg), underwent corrective surgery. All patients were categorized according to surgical age: there were 36 infants aged <1 year, 22 children from 1 to 14 years old, none teenagers from 14 to 18 years of age, and 3 adults more than 18 years of age. Cyanosis was present in 36 patients, respiratory distress and shortness of breath in 22 patients, hemoptysis in 5 patients, and endotracheal intubation and assisted ventilation for cardiopulmonary insufficiency performed in 2 patients. The pulmonary artery systolic pressure (PASP) patients before surgery was 55.3 ± 11.2 mmHg, and pulmonary artery diastolic pressure (PADP) was 29.7 ± 12.5 mmHg, mPAP was 40.3 ± 10.6 mmHg, and mean pulmonary pressure/systemic pressure (Pp/Ps) ratio was 0.61 ± 0.15 (pulmonary artery pressure [PAP] was measured directly by needle before cardiopulmonary bypass [CPB] was established); hemoglobin was 137.8 ± 39.9 g/L and SaO₂ was $76.8 \pm 19.3\%$. Diagnosis was confirmed by physical examination, chest X-ray (cardiothoracic ratio was 0.61 ± 0.06), electrocardiogram, and echocardiography in all patients. Computed tomography scan was performed in 17 patients and right heart catheterization in 9 patients.

According to criteria of the European Association of Pediatric Cardiology, European Association of Cardiothoracic Surgery, and Society of Thoracic Surgery, patients were classified into three DORV types, excluding TOF type. VSD was present in 30 patients, TGA (Taussig-Bing malformation) in 25 patients, and remote VSD in 6 patients. Aortic overriding (AO) of $\geq 90\%$ occurred in 34 patients and AO of 50–89% in 27 patients [Table 1]. The heart in normal position was found in 58 patients, dextrocardia in 2 patients, and mesocardia in

Table 1: Degree of aortic overriding in different types of double outlet right ventricle, *n*

Aortic overriding degree (%)	VSD type	TGA type		Remote VSD type
		Taussig-Bing	TGA	
≥ 90	18	6	4	6
80–89	3	2	0	0
70–79	5	1	1	0
60–69	4	4	0	0
50–59	0	7	0	0

VSD: Ventricular septal defect; TGA: Transposition of the great arteries.

1 patient. Normal great artery relationships were found in 20 patients, side-by-side position in 13 patients, the aorta located to the left anterior in 3 patients, to the right posterior in 7 patients, and to the right anterior in 18 patients. Associated anomalies included atrial septal defect (ASD) in 30 patients, patent ductus arteriosus (PDA) in 33 patients, aortic arch coarctation (CoA) in 6 patients, moderate-to-severe mitral regurgitation in 2 patients, mitral stenosis in 1 patient, and tricuspid valve insufficiency in 1 patient.

Operative technique and operative data

A previous banding procedure was performed in one patient with VSD type, banding + central shunt procedure in one patient with TGA type, and banding + CoA correction in one patient with TGA type.

All operations were performed under general anesthesia with CPB and hypothermia. An intracardiac channel procedure was performed in 37 patients, 8 of whom (revision surgery in one patient with VSD type) underwent right ventricle outflow tract (RVOT) widening using an autologous pericardial patch (two patients with VSD type, four with remote VSD, and one with TGA type). VSD enlargement was performed in 12 patients (two patients with remote VSD, nine with VSD type, and one with multiple VSD). An arterial switch procedure was performed in 19 patients and Rastelli procedure in one patient. To reduce hypoxia, Senning procedure was carried out in three patients and Mustard procedure in one patient. Reoperation was performed in three patients including repair of residual VSD shunts in two patients (remote VSD type) and repair of VSD shunt associated with RVOT widening in one patient (VSD type) [Table 2]. Combined procedures included PDA ligation in 33 patients, ASD repair in 29 patients, mitral valve repair in 3 patients, and tricuspid valve repair in 1 patient.

Statistical analysis

All values were calculated using IBM SPSS software version 19.0 (IBM Corp., Armonk, NY, USA). Continuous variables were expressed as mean \pm standard deviation (SD) or median (IQR), and categorical data were presented as percentages. Student's *t*-test was performed on continuous variables with normal distributions to test the significance of differences, Mann-Whitney *U*-test for variables with nonnormal distributions, and Chi-squared test was used for categorical variables. A value of $P < 0.05$ was considered statistically significant.

RESULTS

Fifty-five (90.2%) patients had an eventual complete recovery. Overall, PAP dropped significantly after corrective surgery (measured directly after CPB was removed); PASP fell from 55.3 ± 11.2 mmHg to 34.7 ± 11.6 mmHg ($t = 14.05$, $P < 0.001$), PADP fell from 29.7 ± 12.5 mmHg to 18.6 ± 7.9 mmHg ($t = 7.39$, $P < 0.001$), mPAP dropped from 40.3 ± 10.6 mmHg to 25.7 ± 8.3 mmHg ($t = 11.85$, $P < 0.001$), and Pp/Ps fell from 0.61 ± 0.15 to 0.39 ± 0.12 ($t = 13.44$, $P < 0.001$) [Table 3]. Six (9.8%) patients died from complications including low cardiac output syndrome in two patients, respiratory failure in two, pulmonary hemorrhage in one, and sudden death in one patient.

Patient outcomes

Infant group

Thirty-one of 36 infants recovered uneventfully, with PASP falling from a preoperative mean of 53.54 ± 10.16 mmHg to 32.88 ± 9.98 mmHg ($t = 12.46$, $P < 0.001$), PADP falling from 26.75 ± 11.35 mmHg to 16.54 ± 7.37 mmHg ($t = 5.3$, $P = 0.001$), Pp/Ps falling from 0.63 ± 0.15 to 0.39 ± 0.12 ($t = 11.64$, $P < 0.001$), and mPAP falling from 38.33 ± 9.42 mmHg to 23.46 ± 7.56 mmHg ($t = 10.47$,

$P < 0.001$). Five infants died owing to low cardiac output syndrome (two patients) and pulmonary complications (three patients), with mortality of 13.9%.

Child group

Twenty-two patients in this group were cured through surgical treatment, with no deaths or further surgery required. PASP in these patients dropped from a preoperative value of 53.93 ± 11.66 mmHg to 37.93 ± 13.56 mmHg ($t = 6.93$, $P < 0.001$), PADP dropped from 33.00 ± 13.20 mmHg to 21.50 ± 8.48 mmHg ($t = 4.57$, $P = 0.002$), Pp/Ps fell from 0.57 ± 0.13 to 0.40 ± 0.13 ($t = 6.91$, $P < 0.001$), and mPAP fell from 41.36 ± 11.78 mmHg to 28.86 ± 9.96 mmHg ($t = 5.72$, $P = 0.001$).

Adult group

Surgical procedures were performed in three adult patients. Two patients underwent Mustard and Senning procedures to reverse the features of hypoxia, and one patient received an intracardiac channel procedure. One patient died (mortality was 33.3%) from low cardiac output syndrome 3 days after the Senning procedure. Overall, PASP dropped from 63.7 ± 10.1 mmHg to 50.3 ± 3.8 mmHg, Pp/Ps fell from 0.61 ± 0.1 to 0.43 ± 0.1 , and mPAP fell from 60.3 ± 9.3 mmHg to 40.7 ± 6.7 mmHg.

Double outlet right ventricle type, surgical procedure, and outcome

Ventricular septal defect type (30 patients)

Correction of VSD type DORV was not difficult when an intracardiac channel procedure was performed, and the results were generally satisfactory. In this group, all patients received an intracardiac channel procedure. However, four patients died at age < 1 year owing to low cardiac output syndrome and pulmonary complications. RVOT stenosis required a secondary operation in one patient.

Transposition of the great arteries type (25 patients)

There were 19 patients with TGA-type DORV whom underwent arterial switch operations in this group. Ages ranged from 15 days to 3 years and 3 months (median: 0.54 years and IQR: 0.33–1.58 years). Only one death owing to sudden death occurred on postoperative day 7. One patient received an intracardiac channel procedure. Senning, Mustard, and Rastelli procedures were performed in five patients to reverse the features of hypoxia; one patient died from low cardiac output syndrome.

Remote ventricular septal defect type (6 patients)

As in the remote VSD type, and for the same reasons, we found that residual shunts could be inadvertently created in the intracardiac channel. Five patients recovered in this group; one patient died of massive pulmonary hemorrhage after revision surgery. Two patients required a secondary operation to repair residual VSD shunts.

Degree of aortic overriding

DORV is associated with varying degrees of AO; however, operative mortality did not appear to be related to this. In our

Table 2: Surgical method selection with different types of double outlet right ventricle

Operation selection	DORV type, <i>n</i>		
	VSD	TGA	Remote VSD
Operation type			
Corrective surgery (intracardiac channel)	30	1	6
Switch	0	19	0
Senning	0	3	0
Mustard	0	1	0
Rastelli	0	1	0
Previous operation			
Banding	1	0	0
Banding + shunt	0	1	0
Banding + correction of CoA	0	1	0
Secondary operation			
RRS	0	0	2
RRS + ERVOTO	1	0	0

DORV: Double outlet right ventricle; CoA: Coarctation of aorta; RRS: Repair of VSD residual shunt; ERVOTO: Enlargement of right ventricle outlet tract obstruction; VSD: Ventricular septal defect; TGA: Transposition of the great arteries.

Table 3: PAP values before and after surgery (*n* = 61)

Variables	Preoperation	Postoperation	<i>t</i>	<i>P</i>
PASP (mmHg)	55.3 ± 11.2	34.7 ± 11.6	14.05	< 0.001
PADP (mmHg)	29.7 ± 12.5	18.6 ± 7.9	7.39	< 0.001
mPAP (mmHg)	40.3 ± 10.6	25.7 ± 8.3	11.85	< 0.001
Pp/Ps	0.61 ± 0.15	0.39 ± 0.12	13.44	< 0.001

Data are shown as mean \pm SD. PASP: Pulmonary artery systolic pressure; PADP: Pulmonary artery diastolic pressure; mPAP: Mean pulmonary arterial pressure; Pp/Ps: Pulmonary pressure/systemic pressure; SD: Standard deviation; PAP: Pulmonary artery pressure.

study, three of 27 patients died in the group with 50–89% right-sided AO. Another three patients died in the group of 34 patients with over 90% AO. There was no significant difference in mortality between these two groups ($\kappa = 0.09$, $P = 1.00$).

DISCUSSION

Except for TOF type, which usually has lower PAP, DORV is a group of complex congenital heart abnormalities that is initially associated with excessive pulmonary blood flow and frequent recurrent pulmonary infections.^[10] Cyanosis may be present soon after birth. The onset of PH often leads to a progressive reduction in exercise capacity and fatigue resulting from impaired respiratory and heart function, which cause stunted childhood development. The disorder is complex and is often associated with other forms of intracardiac or extracardiac malformation. PH usually arises early and eventually results in heart and lung dysfunction, with a missed opportunity to performing surgery. Therefore, once the diagnosis is confirmed, surgical treatment should be considered. In addition, we should give special consideration on the following aspects of management.

Timing of operation and indications

It is generally considered that these patients often develop significant congestive heart failure due to a large left-to-right shunt, and surgical repair should be carried out in the first several months of life.^[11] In our series, 31 of 36 patients aged <1 year recovered uneventfully. However, 5 of 36 infants died (13.9%), 3 of the 36 infant patients underwent secondary surgery to correct postoperative complications (two patients underwent successful closure of a residual shunt in the intracardiac channel, and one patient was revised for a residual shunt in the intracardiac channel combined with enlarging an RVOT stenosis). In contrast to the infant group above, all operations were successfully performed in the 22 patients aged between 1 and 14 years (median: 2.00 years and IQR: 1.50–4.30 years), including in 3 patients who had undergone a previous banding procedure. No patients died or required further surgery. Senning procedure and Mustard procedure were performed in three adult patients to reverse the features of hypoxia; one died from low cardiac output syndrome. Although the average survival time after diagnosis of PH was estimated to be 5–7 years nowadays, the mortality was raised significantly after 3 years old.^[12–15] Therefore, according to our observation, it was safe for patients with DORV and PH to receive surgical treatment between 1 and 3 years old. Younger infants and adult patients may have higher mortality.

Correction of VSD type DORV is not difficult when an intracardiac channel procedure is performed, with generally good results.^[9] In infants <1 year old, however, we found that residual shunts could be inadvertently created in the intracardiac channel, especially for DORV with remote VSD. This could be owing to more difficult exposure of the surgical field, myocardial fragility, and difficulty in suturing. RVOT stenosis required a secondary operation. There were 19 patients with TGA type DORV in the group

who underwent arterial switch operations, with only one sudden death on the 7th postoperative day. Outcomes of the arterial switch procedure were generally excellent. In our experience, surgical treatment is difficult in patients with remote VSD type DORV, and mortality is a concern. In our group of six patients, five recovered and one patient died of massive pulmonary hemorrhage after revision surgery.

Overall, infants and children in this study had a significant reduction in PH after surgery. However, the decline was not significant for a few infants <1 year old, which may be related to hypoplastic development of the pulmonary vascular bed. Serious pathological changes are caused by excessive pulmonary blood flow, leading to hypertension in the pulmonary vascular tree.^[16] In this study, PAP could not be significantly reduced in three adults; the purpose of surgery was to increase systemic blood oxygen saturation by influencing intracardiac streaming, which can reduce symptoms and improve the quality of life through intra-atrial bypass as part of a palliative care intervention.^[17] However, operative risk remains high in these patients.

Effect of associated lesions on operative outcomes

VSD type of DORV is usually associated with PH; the operative results are acceptable in this group. Our results suggested that surgery should be performed between ages of 1 and 3 years to obtain the most satisfactory results; PAP can be significantly reduced with uneventful recovery.

However, the operative treatment of DORV with remote VSD is more difficult. Cure remains feasible, but the condition must be managed individually. DORV can be associated with abnormal location and spatial relationship of the main arteries, or the presence of excessive conal muscle, subarterial fibrous tissue, and other cardiac malformations. The two great arteries may be parallel or located in an anterior and posterior position, or the aorta right or left anterior to the pulmonary artery trunk. Owing to the positional anomalies of these arteries and their abnormal relationship with the VSD, the pathology is more complex and surgical correction more difficult, resulting in greater operative risk among these patients.

All our patients under 3 years of age (including infants) with TGA type DORV underwent an arterial switch procedure, with satisfactory results. We found that the reduction in PAP after surgery might not be significant in patients with TGA older than 3 years, the same trend that also increased in adult patients. The surgery risk is quite significant, even for palliative surgery.

Choice of surgical method and prevention of complications

Pathological changes in patients with DORV are varied, necessitating an individualized surgical strategy to optimize success. The suture technique used in intracardiac channel repair of VSD type DORV should be sufficient to prevent residual shunt formation. In cases of restricted VSD, enlargement of the outflow defect should be carried out, with attention paid to avoid outflow tract stenosis from both the left and right ventricular chambers. To avoid RVOT

stenosis, the tract should simultaneously be enlarged with a patch. Eight of our patients in this group received RVOT patch widening. For patients with DORV of remote VSD type, a total intracardiac conduit may be needed. For TGA type DORV, an arterial switch operation remains the best choice. During the operation, coronary anastomotic patency should be confirmed, and care taken to prevent distortion and stenosis. Aortic and pulmonary valves should be protected. It is important to avoid stenosis of the pulmonary artery and its branches during pulmonary artery anastomosis. If the aorta and pulmonary artery are located in an anterior and posterior position and the VSD is below the pulmonary valve, arterial switch surgery can be performed after establishing an intracardiac channel.

Mustard or Senning surgical techniques are generally safe because of their relative simplicity. Both methods are useful for neglected cases with severe PH, cyanosis, and hypoxia, in which the best opportunity for surgery has been missed. These palliative procedures will improve hypoxia and overall quality of life; however, the operative risk is high. During surgery, attention should be paid to prevent lung injury and maintain cardiac function; consequently, VSD closure can be avoided. One patient with mild PH (<50 mmHg) was deemed suitable to undergo Rastelli surgery, with careful paid to prevent distortion and stenosis of intracardiac and extracardiac channels that would normally require reoperation. DORV is often associated with other intracardiac malformations such as ASD, PDA, and atrioventricular valve regurgitation, which require concurrent correction.

Postoperative management

The postoperative management of these patients is very important. Routine use of inhaled nitric oxide and prostaglandins is helpful. Diuretic drugs and other supportive agents are essential. Sedation and adequate respiratory support, including extended ventilation, should be emphasized.

In conclusion, owing to the wide variety of pathologies encountered in DORV, the operative outcome is dependent on several factors including patient age, type of lesion, associated malformations, PH, and surgical technique. Most patients can achieve a satisfactory surgical outcome with an individualized surgical strategy and technique and proper postoperative management.

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Conflicts of interest

There are no conflicts of interest.

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