Impact of anatomic characteristics and initial biventricular surgical strategy on outcomes in various forms of double-outlet right ventricle

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ABSTRACT

Objectives: Surgical management of various forms of double-outlet right ventricle uses a variety of approaches depending on the underlying anatomic form. In this study, we sought to determine the risk factors of mortality and reoperation in those with double-outlet right ventricle undergoing biventricular repair, according to anatomic characteristics and initial surgical strategy.

Methods: Between 1992 and 2013, 433 patients were included in the study. Double-outlet right ventricle was classified as double-outlet right ventricle with subaortic ventricular septal defect associated with subpulmonary obstruction in 33% of patients (n = 141), with subaortic ventricular septal defect without subpulmonary obstruction in 30% of patients (n = 130), with subpulmonary ventricular septal defect in 32% of patients (n = 139), and with noncommitted ventricular septal defect in 5% of patients (n = 23). Three types of repairs were performed: (1) intraventricular baffle repair, n = 149 (34%); (2) intraventricular baffle repair with right ventricular outflow tract reconstruction, n = 163 (38%); and (3) intraventricular baffle repair with arterial switch operation, n = 121 (28%).

Results: Thirty-day overall mortality was 7.4%. Early reoperation was needed in 6% of the cases. Early mortality was higher in the intraventricular baffle repair with arterial switch operation group ($P = .01$). Survival at 10 years was 86.2%, and freedom from reoperation at 10 years was 61.4%. At last follow-up (median, 5.7 years; 95% confidence interval, 4.5-6.6), mortality and reoperation rates were similar in the different surgical strategy groups. Late reoperation and late mortality were significantly higher in the double-outlet right ventricle with noncommitted ventricular septal defect group ($P < .01$). In multivariate analyses, risk factors for reoperation were concomitant surgical procedures ($P = .03$) and duration of cardiopulmonary bypass ($P < .01$). Risk factors for mortality were restrictive ventricular septal defect ($P = .01$), mitral cleft ($P < .01$), and associated coronary artery anomalies ($P = .01$).

Conclusions: Those with the anatomic type of double-outlet right ventricle with noncommitted ventricular septal defect were at higher risk for reoperation and mortality. Intraventricular baffle repair with arterial switch operation was the surgical strategy in patients at higher risk of early death. Initial surgical strategy did not influence the late outcomes. (J Thorac Cardiovasc Surg 2016;152:698-706)

Freedom from reoperation according to surgical strategy.

Central Message

The initial surgical strategy did not influence late outcomes of DORV with biventricular repair.

Perspective

The outcomes of DORV with biventricular repair remain unclear. Between 1992 and 2013, 433 consecutive patients were retrospectively included. Early mortality was higher in the IVR-ASO group. Late mortality and late reoperation rates were similar in the different biventricular surgical strategy groups, but were significantly higher in DORV with ncVSD. Multiple risk factors have been identified.

See Editorial Commentary page 707.

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Received for publication Dec 12, 2015; revisions received April 26, 2016; accepted for publication May 9, 2016; available ahead of print June 24, 2016.

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0022-5223/\$36.00

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<http://dx.doi.org/10.1016/j.jtcvs.2016.05.019>

Abbreviations and Acronyms

ASO	= arterial switch operation
AV	= atrioventricular
AVSD	= atrioventricular septal defect
CI	= confidence interval
CPB	= cardiopulmonary bypass
DORV	= double-outlet right ventricle
IVR	= intraventricular baffle repair
ncVSD	= noncommitted ventricular septal defect
OR	= odds ratio
REV	= réparation à l'étage ventriculaire
RVOT	= right ventricular outflow tract
VSD	= ventricular septal defect

Scanning this QR code will take you a video and supplemental tables.

Double-outlet right ventricle (DORV) is a type of ventriculoarterial connection (or alignment) of the great arteries in which both great arteries arise entirely or predominantly from the right ventricle.¹ DORV is a heterogeneous group of congenital heart diseases subclassified by the International Nomenclature Committee for Pediatric and Congenital Heart Disease, according to the relationship of the ventricular septal defect (VSD) to the great arteries and to the presence or absence of pulmonary outflow tract obstruction (International Pediatric and Congenital Cardiac Code).²

In 1964, Kirklin and colleagues³ reported a successful biventricular repair in a child with DORV, subaortic VSD, and concordant atrioventricular (AV) connections. Since then, complete correction through the use of a variety of surgical techniques has been achieved in more complex forms of DORV. The feasibility of a biventricular repair still remains a surgical challenge in the management of DORV.^{4,5} Previous series reported successful biventricular surgical repair in different anatomic subtypes of DORV.⁶⁻⁸ Thus far, however, 2 points still remain unclear: which surgical strategy implies a higher risk of mortality or reoperation and when to abandon the ambition of biventricular repair. In this study, we sought to determine the risk factors for mortality and reoperation in those with DORV undergoing biventricular repair according to anatomic characteristics and initial surgical strategy.

MATERIALS AND METHODS

Patients

We reviewed all records of consecutive patients with DORV who underwent operation in 2 French centers (Necker Enfants Malades, Paris,

France; Centre Chirurgical Marie Lannelongue, Le Plessis Robinson, France) between 1992 and 2013. Those with conotruncal anomalies with the presence of double conus but without both great arteries arising entirely or predominantly from the right ventricle⁹ were not included in this cohort. Those with DORV associated with double-inlet ventricle, absent AV connection, or severely imbalanced ventricles precluding biventricular repair were excluded. The diagnosis of DORV with 2 balanced ventricles (or mild asymmetry not precluding biventricular repair) was confirmed after review of echocardiograms, cardiac magnetic resonance imaging or computed tomography scans, angiograms, and surgical reports. In all cases, the type of DORV was finally confirmed according to the anatomic description given by the surgeon. When biventricular repair was considered unattainable in light of both preoperative investigations or intraoperative evaluation, the patient was excluded. Demographic data included gender, prenatal or postnatal diagnosis, center, and associated genetic anomalies.

Four anatomic groups were defined according to the classification of the International Pediatric and Congenital Cardiac Code²: (1) subaortic or doubly committed VSD and subpulmonary obstruction, Fallot type; (2) subaortic or doubly committed VSD without subpulmonary obstruction, VSD type; (3) subpulmonary VSD, Taussig-Bing type¹⁰; and (4) noncommitted VSD (ncVSD) type.

Along with Lev and colleagues,¹¹ we defined the VSD in DORV according to the direction of the blood flow coming from the left ventricle through the VSD: directly into the aorta (subaortic VSD), the pulmonary artery (subpulmonary VSD), or both (doubly committed VSD). These "committed" VSDs always open in the outlet of the right ventricle and are cradled within the limbs of the septal band (outlet VSD). Conversely, the blood flow through an ncVSD is not directed toward the outflow tract, but directly toward the right ventricular cavity, running perpendicular to the ventricular septum. We include in this category all the VSDs that are not located between the 2 limbs of the septal band, that is inlet, muscular, and central membranous VSD.¹²

Associated cardiac anomalies were noted: VSD (location, number, and restriction when significant pressure gradient between the ventricles was diagnosed), subpulmonary and subaortic obstruction, atrioventricular septal defect (AVSD), mitral valve cleft and other mitral valve anomalies, straddling of the mitral or of the tricuspid valve, and coronary artery anomalies.

Three surgical strategies were used: (1) intraventricular baffle repair (IVR); (2) IVR with right ventricular outflow tract (RVOT) reconstruction, including IVR with RVOT patch enlargement, réparation à l'étage ventriculaire (REV), and Rastelli procedures, IVR-RVOT; and (3) IVR with arterial switch operation (ASO), IVR-ASO.

Concomitant surgical procedures were reported: VSD enlargement, AV valve repair, correction of abnormal pulmonary venous return, coarctation or interrupted aortic arch repair, chordae mobilization, subaortic stenosis resection, and pulmonary artery branch stenosis repair.

Surgical data included prior palliative intervention, age and weight at time of repair, date of surgery, surgical strategy, concomitant surgical procedure, cardiopulmonary bypass (CPB) duration, duration of mechanical ventilation, reoperation, and delay between repair and reoperation.

Finally, we collected early mortality and reoperation occurring less than 30 days after repair and late mortality and reoperation occurring more than 30 days after repair. Follow-up information was available in 341 of 381 surviving patients (90%). This study was approved by our ethics board (Comité de Protection des Personnes-Ile-de-France VI).

Statistical Analysis

Continuous variables are presented as the mean value $\pm$ standard deviation when variables were normally distributed and median value with 95% confidence interval (CI) when they were not. Categorical variables are presented as percentages. Comparisons of categorical variables were made using the chi-square test or Fisher exact test when appropriate.

Time to death and time to reoperation are shown in Kaplan–Meier curves and all actuarial curves start at the time of surgery. Cox proportional hazard regression analysis, with the date of repair used as the start date, was performed as univariate and multivariate analyses (Table E1 shows a list of variables considered for multivariable analyses) to investigate the risk factors for early death, late death, early reoperation, and late reoperation. When there were less than 10 outcome events per predictor variable, we used the chi-square or Mann–Whitney test to determine it as appropriate. We excluded early death and reoperation from the analysis when we evaluated late death or reoperation. All entered variables were selected on the basis of clinical experience, univariate analysis, and previously published data. Receiver operating curve analysis was used to study the ability of significant parameters to predict early death. Analysis was conducted using MedCalc (MedCalc Software, Mariakerke, Belgium).

RESULTS

Population and Anatomic Characteristics

Overall, we identified 482 patients with DORV. Eighteen patients had only a palliative procedure (death before biventricular repair) and were excluded. Thirty-one patients (6%) followed a sequential cavopulmonary connection program and were excluded: 12 with DORV with ncVSD, 9 with multiple VSD, 5 with straddling mitral valves, and 5 with other severe mitral valve anomalies.

Finally, 433 DORV cases with 2 adequate ventricles underwent biventricular repair and were included in the study. Demographic and anatomic characteristics are summarized in Table 1. The surgical procedures are shown in the Video 1 (DORV with ncVSD with IVR-ASO).

The number and type of prior palliative interventions according to the anatomic groups are shown in Table 2. The 18 interventions listed in the group “Others” included 6 isolated pulmonary artery branch repair, 4 closures of the arterial duct, 3 partial cavopulmonary connections (converted secondarily to biventricular strategy), 2 corrections of partial anomalous pulmonary venous return, 1 tricuspid valve repair, 1 aberrant left pulmonary artery branch reimplantation, and 1 pacemaker implantation. The proportion of DORV with ncVSD requiring a palliative procedure before repair (12/23, 52%) was significantly higher than in the VSD type (41/130, 32%; $P < .01$), Fallot type (44/141, 31%; $P < .01$), and Taussig–Bing type (38/139, 27%; $P < .01$). Table 3 shows the surgical strategy in the 4 anatomic groups with concomitant surgical procedures.

Survival and Reoperation

The median follow-up was 5.7 years (95% CI, 4.5–6.6). The overall survival was 87.8% at 1 year, 87.2% at 5 years, and 86.2% at 10 years. The overall freedom from reoperation rate was 86.5% at 1 year, 74.1% at 5 years, and 61.4% at 10 years. At last follow-up, 351 of 381 survivors (92%) were in New York Heart Association functional class I, and 30 of 381 survivors (8%) were in New York Heart Association functional class II.

TABLE 1. Patient characteristics

Variable	No. (%)
Demographic characteristics	
Male/female	260 (60)/173 (40)
Center (Necker/Plessis Robinson)	216 (50)/217 (50)
Prenatal diagnosis	171 (39)
Chromosomal/genetic abnormalities	50 (12)
IPCCC classification	
Fallot type	141 (33)
VSD type	130 (30)
Taussig–Bing type	139 (32)
ncVSD type	23 (5)
Anatomic characteristics	
VSD	
No. of VSDs	
1	373 (86)
Multiple	61 (14)
Restrictive VSD	35 (8)
Ventriculoarterial spatial relationship	
D-malposition	343 (79)
D side-by-side	42 (10)
L-malposition	24 (5.5)
L side-by-side	2 (0.5)
Undetermined	22 (5)
Subpulmonary obstruction	
Pulmonary atresia	19 (4)
Stenosis (infundibular/valvular/supravulvular)	165 (38)
Subaortic and supra aortic obstructions	
Valvular or subvalvular aortic stenosis	21 (5)
Coarctation of the aorta	62 (14)
Interrupted aortic arch	18 (4)
AV valve anomalies	
AVSD	12 (3)
Straddling	67 (16)
Mitral/tricuspid/bivalvular	17 (4)/42 (10)/8 (2)
Other mitral valve anomalies	36 (8)
Coronary artery anomalies	
DORV Taussig–Bing type	112 (26)
	67/139 (48)

IPCCC, International Pediatric and Congenital Cardiac Code; VSD, ventricular septal defect; ncVSD, noncommitted ventricular septal defect; AV, atrioventricular; AVSD, atrioventricular septal defect; DORV, double-outlet right ventricle.

Outcomes according to double-outlet right ventricle type. Those with noncommitted VSD had a higher risk of mortality ($P = .02$) (Figure 1, A) and reoperation ($P = .04$) (Figure 1, B) in comparison with those with other anatomic types. Mortality was higher (hazard ratio, 2.18; 95% CI, 1.10–4.30) and reoperation was more frequent (hazard ratio, 1.68; 95% CI, 1.07–2.63) in the Taussig–Bing type than in the VSD type. No difference was found in mortality or reoperation rate between the Taussig–Bing type and Fallot type, and between the VSD type and the Fallot type.

Outcomes according to surgical strategy. No significant statistical difference was found in late mortality ($P = .20$) (Figure 2, A) and late reoperation rate ($P = .13$) (Figure 2, B) according to the 3 surgical strategies.

VIDEO 1. Example of IVR + ASO on a DORV with ncVSD + restrictive VSD. Video available at: [http://www.jtcvsonline.org/article/S0022-5223\(16\)30457-3/addons](http://www.jtcvsonline.org/article/S0022-5223(16)30457-3/addons).

Early mortality. A total of 32 of 433 patients died within 30 days after repair (7.4%). The causes of death were as follows: 12 during surgery (5 during IVR + ASO, with no significant statistical difference compared with the other surgical strategies; odds ratio [OR], 1.7; 95% CI, 0.4-5.7; $P = .47$), 4 during IVR with concomitant surgical procedure (AV valve repairs and coarctation repairs), and 3 during IVR-RVOT with concomitant surgical procedures (AV valve repairs, correction of APVR, and subaortic stenosis resection); 7 sepsis cases, 4 low cardiac output syndromes, 4 pulmonary hypertension crises, 2 electromechanical dissociations, 1 hemorrhage, and 2 unclearly explained. Early postoperative characteristics are shown in Table 4. The IVR + ASO was the surgical strategy with the highest rate of early death (OR, 3.1; 95% CI, 1.4-7.1; $P = .01$). The causes of early death after IVR + ASO (15 patients) were 5 during surgery, 4 sepsis cases, 2 low output cardiac failures, 1 pulmonary hypertension crisis, 1 electromechanical dissociation, 1 hemorrhage, and 1 unclearly explained. Early mortality was significantly higher in DORV with ncVSD ($P = .04$), low weight at operation ($P = .04$), left ventricular outflow tract obstruction ($P = .04$), and associated surgical procedure ($P = .03$). By using receiver operating characteristics, weight less than 4000 g predicted

TABLE 2. Prior palliative interventions according to anatomic groups (180 procedures in 135 patients [31%])

	Fallot type	VSD type	Taussig-Bing type	ncVSD type	Total
No. of patients	44	41	38	12	135
PA banding	2	25	10	12	49
PA banding + CoA repair	0	16	10	1	27
Blalock shunts	42	0	14	15	71
Atrial septectomy	2	1	8	4	15
Others	6	2	6	4	18
Total	52	44	48	36	180

VSD, Ventricular septal defect; ncVSD, noncommitted ventricular septal defect; PA, pulmonary artery; CoA, coarctation of the aorta.

TABLE 3. Surgical strategy according to anatomic group

	Fallot type	VSD type	Taussig-Bing type	ncVSD type	Total
IVR	28	114	0	7	149
IVR-RVOT	113	16	23	11	163
REV	2	0	9	0	11
Rastelli	11	1	12	5	29
IVR-ASO	0	0	116	5	121
Total	141	130	139	23	433
Concomitant surgical procedures					
Enlargements of restrictive VSD	11	15	0	9	35
Coarctation repairs	0	13	16	3	32
AV valve repairs	13	6	3	2	24
Subaortic stenosis resections	2	6	5	3	16
APVR repairs	2	2	0	2	6
Pulmonary artery branch plasties	2	2	1	0	5
Chordae mobilizations	1	0	0	3	4
Total	31	44	25	22	122

VSD, Ventricular septal defect; ncVSD, noncommitted ventricular septal defect; IVR, intraventricular repair; RVOT, right ventricular outflow tract; REV, réparation à l'étage ventriculaire; ASO, arterial switch operation; AV, atrioventricular; APVR, abnormal pulmonary venous return.

early mortality with a sensitivity of 80% and a specificity of 68%.

Early reoperation. Early cardiac reoperation was needed in 26 patients (6%) with no significant statistical differences between the anatomic types of DORV and the surgical strategies. The most frequent cause of early reoperation for subaortic VSD (Fallot type and VSD type) was residual VSD (35%, 9/26) (Table 5).

Late mortality. A total of 20 patients died, 16 (80%) within the first year after repair. Causes of death were end-stage heart failure in 8 patients, complications of a mechanical valve in 3 patients, sudden death in 3 patients, death after reoperation in 2 patients (1 valve replacement after endocarditis, 1 RVOT enlargement), noncardiac death in 2 patients, and 1 missing information. No significant difference in late mortality was found between the anatomic types or the surgical strategies. Factors associated with late mortality for each surgical strategy in univariate analysis are shown in Table E2. In multivariate analysis, risk factors for late mortality were IVR: mitral valve abnormality ($P = .01$) and concomitant procedure ($P < .01$); IVR-RVOT reconstruction: subaortic obstruction ($P = .02$); and IVR-ASO: concomitant procedure ($P < .01$).

In the overall analysis, the factors associated with late mortality in univariate analysis are shown in Table E3. To determine the anatomic characteristics that affected late mortality, we performed a multivariate backward analysis (Table E4) including restrictive VSD, mitral cleft, and coronary artery anomalies. Risk factors for late mortality were restrictive VSD (OR, 2.7; 95% CI, 1.2-6.3; $P = .01$),

FIGURE 1. Outcomes according to anatomic type of DORV. A, Survival according to anatomic type of DORV. B, Freedom from reoperation according to anatomic type of DORV. CI, Confidence interval; VSD, ventricular septal defect.

mitral cleft (OR, 3.5; 95% CI, 1.6-5.6; $P < .01$), and coronary artery anomalies (OR, 2.9; 95% CI, 1.5-3.9; $P = .01$).

Late reoperation. A total of 97 patients underwent 136 reoperations at a median of 3.1 years after repair (95% CI, 0.1-13) (Tables 6 and 7). A total of 71 patients underwent only 1 reoperation, 16 patients underwent 2 reoperations, 8 patients underwent 3 reoperations, 1 patient underwent 4 reoperations, and 1 patient underwent 5 reoperations. Freedom from reoperation was not different according to anatomic group or surgical strategy. Factors associated with late reoperation for each surgical strategy in univariate analysis are shown in Table E5. In multivariate analysis, risk factors for late reoperation were IVR: restrictive VSD ($P = .04$) and subaortic obstruction ($P = .04$); IVR-RVOT reconstruction: duration of CPB

FIGURE 2. Outcomes according to surgical strategy. A, Survival according to surgical strategy. B, Freedom from reoperation according to surgical strategy. CI, Confidence interval; IVR, intraventricular baffle repair; ASO, arterial switch operation; RVOT, right ventricular outflow tract.

($P < .01$); and IVR-ASO: subaortic obstruction ($P = .03$), concomitant procedure ($P < .01$), and days of intubation ($P < .01$).

In the overall analysis, risk factors associated with reoperation in univariate analysis are shown in Table E3. In multivariate analysis (Table E4), including IPCC classification, surgical strategy, concomitant surgical procedure, and duration of CPB, concomitant surgical procedure (OR, 1.6; 95% CI, 1-2.5; $P = .03$) and duration of CPB (OR, 1.004; 95% CI, 1.004-1.009; $P < .01$) were the 2 factors associated with late reoperations.

DISCUSSION

We report a large series of patients with DORV who underwent biventricular repair over a period of 21 years

TABLE 4. Early surgical and postoperative characteristics (<30 days) according to surgical strategies

	IVR (n = 149)	IVR-RVOT reconstruction (n = 163)	IVR-ASO (n = 121)	P value
Surgery				
Age at repair (d, median [95% CI])	147 [121-235]	423 [318-597]	26 [18-57]*	<.01
Weight (kg, median [95% CI])	5.2 [4.6-6.0]	7.9 [7.0-8.6]	3.4 [3.3-3.5]*	.02
Duration of CPB (min, mean)	117	140	198	.63
Aortic clamping time (min, mean)	71	81	121	.41
Concomitant procedures	42	34	39	.29
Postoperative characteristics				
Inotropic support (d, median [95% CI])	3 [2-4]	3 [2-5]	7 [6-9]*	.01
Duration of intubation (d, median [95% CI])	4 [3-5]	4 [3-5]	8 [7-10]*	.03
% Reoperation (<30 d)	4.7%	5.5%	8.3%	.09
% Mortality (<30 d)	7%	4.3%	10.7%*	.01

IVR, Intraventricular repair; RVOT, right ventricular outflow tract; ASO, arterial switch operation; CI, confidence interval; CPB, cardiopulmonary bypass. *P < .05.

in 2 surgical centers. The early mortality rate was 7.4%, and 10-year overall mortality rate was 13.8%. The reoperation rate was 6% for early reoperation, and 10-year overall reoperation rate was 38.6%. Anatomic type of DORV influenced the outcomes, because ncVSD was associated with a higher risk of death and reoperation compared with the other groups. Mortality was higher in the Taussig–Bing type than in the VSD type. These differences may be related to the fact that the anatomic type of DORV is the main determinant of the surgical strategy. However, late survival and freedom from reoperation were not different for the 3 surgical cohorts.

Li and colleagues⁸ recently reported a similar series with a shorter follow-up, with early death in 4.4%. The contemporary character of the study and lower proportion of Taussig–Bing type (20%) in their series may explain the difference with the present report. Kleinert and colleagues⁵ reported early death in 8% (12/148 DORV with biventricular repair) with a proportion of anatomic subtypes identical to our series.

Outcomes According to Anatomic Characteristics

The proportion of the different types of DORV in our series is similar to the majority of previously reported

series,^{5,6} with 60% of subaortic (or doubly committed) VSD, 30% to 35% of subpulmonary VSD¹³ (ie, Taussig–Bing heart), and 5% to 10% of ncVSD.¹⁴

We observed that DORV with ncVSD was the only anatomic subgroup in which early and late mortality rates were significantly higher compared with other anatomic groups. Belli and colleagues¹⁴ and Lacour-Gayet et al¹⁵ showed the same trends in their series of DORV with ncVSD. Although Brown and colleagues¹⁶ reported biventricular repair in a high proportion of DORV with ncVSD, the choice of this approach as opposed to a univentricular palliation program should be carefully analyzed, because the risk of death and reoperation remains high. The distance between the VSD and the great arteries,¹⁴ the location of the VSD (inlet, muscular, or central perimembranous),¹² and the evaluation of the outflow tracts, AV valves, and global anatomy are the main parameters that should be analyzed to indicate a biventricular repair or a univentricular strategy in DORV with ncVSD. Redirecting the ncVSD to the unobstructed RVOT associated with the ASO procedure, performed in 5 cases in our series, appeared to be a valuable alternative in this unfavorable anatomic subset.

Acquired RVOT obstruction was the main cause of reoperation in the Taussig–Bing type compared with the VSD type. Hayes and colleagues¹³ reported that it was the main cause of reoperation (5/11, 45%) in a series of 43 Taussig–Bing anomalies. In the series by Kleinert and colleagues,⁵ hemodynamically significant subaortic obstruction (valvar or subvalvar aortic stenosis) and anatomic substrate for subaortic obstruction (expressing itself as coarctation of the aorta or interrupted aortic arch) were the only anatomic factors, with VSD location associated with early mortality.

The anatomic factors associated with late mortality (except ncVSD) were restrictive VSD, mitral cleft, and coronary artery anomalies. Restrictive VSD is a major anatomic parameter that influences preoperative clinical tolerance, timing of surgical intervention, duration of procedure, and intracardiac tunnel geometry. The enlargement of

TABLE 5. Indications for early reoperation according to anatomic group

Indication	Fallot type	VSD type	Taussig–Bing type	ncVSD	Total
Residual VSD	4	3	1	1	9
RVOT reconstruction	4		4		8
Coronary artery complications	1		3		4
Subaortic obstruction		1	2		3
Banding of PA		1			1
Mechanical valve thrombosis			1		1
	9	5	11	1	26

VSD, Ventricular septal defect; ncVSD, noncommitted ventricular septal defect; RVOT, right ventricular outflow tract; PA, pulmonary artery.

TABLE 6. Indications for late reoperation according to anatomic group

Indication	Fallot type	VSD type	Taussig–Bing type	ncVSD	Total
Subaortic obstruction	8	15	7	5	35
Aortic valve repair	1	2	10		13
Aortic arch repair			11		11
RVOT reconstruction	13	7	15	3	38
Pulmonary artery branch repair	5		1		6
Residual VSD closure	4	6	4		14
AV valve repair	2		1	2	5
Pacemaker implantation	4	3	2	1	10
Endocarditis		1	1		2
ASD closure			1		1
Heart transplantation			1		1
	37	34	54	11	136

VSD, Ventricular septal defect; ncVSD, noncommitted ventricular septal defect; RVOT, right ventricular outflow tract; AV, atrioventricular; ASD, atrial septal defect.

VSD, which was indicated in front of a diagnostic of restrictive VSD during the preoperative evaluation or the surgical procedure (when the surgeon deemed it necessary to intraventricular baffle), was the most frequent additional procedure (23/45, 51%). In the series by Li and colleagues,⁸ the most frequent additional procedure (23/45, 51%) was the enlargement of the VSD, which was indicated when restrictive VSD was diagnosed during the preoperative evaluation or during the surgical procedure (when the surgeon considered it necessary). This was performed in only 35 of 122 patients (29%) in our series. Nevertheless, the IVR, with or without enlargement of VSD, carries a high risk for reoperation for VSD residual (or modification of baffle repair) ahead of DORVs or all types of conotruncal abnormalities.¹⁷ We believe this surgical parameter requires a particularly precise surgical management (size and localization of septal resection) to avoid risks in the short

term (conduction disorders, residual VSD) and long term (subaortic obstruction). In regard to reoperation for left ventricular outflow obstruction, one may conclude that an aggressive approach with almost routine VSD enlargement may decrease late left ventricular outflow obstruction. Nevertheless, because it was difficult for us to demonstrate this approach statistically, we have to consider these conclusions and suggestions cautiously. A prospective study would be appropriate to investigate this.

Outcomes According to Surgical Strategy

IVR-ASO was the surgical strategy that carried the highest risk of early mortality independently of the anatomic subtype of DORV. Of note, IVR-ASO was performed primarily in infants with lower weight and who required higher duration of CPB (Table 4). These factors are known to be risk factors for early mortality after surgery for congenital heart diseases.^{18,19} The early mortality for IVR-ASO was higher than after the arterial switch for transposition of the great arteries (10.7% vs 6%).²⁰ The need to build an intracardiac baffle from the VSD to the neo-aorta, the higher incidence of abnormal coronary artery origin/course, and the frequent association of aortic arch obstruction, resulting in a longer procedure, may explain this trend.²¹

In this study, we could not demonstrate any differences in late mortality according to the initial strategy. The small number of deaths in the first year postsurgery (4/433, 1%) certainly limits this conclusion. Li and colleagues⁸ reported no death after the first year, but their follow-up was shorter (3.4 ± 3.9 years). Belli and colleagues⁶ reported 2 deaths (2/151, 1%) in their series, with a follow-up of 4.3 years. Late mortality is low after biventricular repair in DORV, and adverse events are mainly reoperations. Indeed, approximately one third of the patients in our series had to undergo a reoperation (Table 7). Some 43% of these reoperations were related to left heart residual or acquired anomalies. The types of left heart anomalies observed were different according to the initial surgical strategy. Subaortic obstruction related to baffle stenosis was the main cause of reoperations in the IVR and IVR-RVOT groups (90% of indications), whereas it occurred in only one third of patients undergoing reoperation after IVR-ASO. Conversely, aortic arch repair or ascending aorta and aortic valve repair were frequent in the IVR-ASO group and rare in the 2 other surgical strategies. These results are similar to those of Losay and colleagues,²² who reported that aortic arch reconstruction (15/39, 38%) and aortic valve repair (13/39, 33%) were the major indications of late reoperation for left heart anomalies in a large series of 1200 transpositions of the great arteries after ASO. Belli and colleagues²³ also showed that subaortic valve obstruction developed with time and was the main cause of reoperation (73%) after IVR or IVR-RVOT. Of note, abnormal coronary artery

TABLE 7. Indications for late reoperation and surgical strategy

Indication	IVR	IVR-RVOT	IVR-ASO	Total
Subaortic obstruction	17	9	9	35
Aortic valve repair	2	1	10	13
Aortic arch repair			11	11
RVOT reconstruction	5	24	9	38
Pulmonary artery branch repair	1	4	1	6
Residual VSD closure	5	3	6	14
AV valve repair	3	1	1	5
Pacemaker implantation	3	4	3	10
Endocarditis	1		1	2
ASD closure			1	1
Heart transplantation		1		1
	37	47	51	136

IVR, Intraventricular repair; RVOT, right ventricular outflow tract; ASO, arterial switch operation; VSD, ventricular septal defect; AV, atrioventricular; ASD, atrial septal defect.

distribution was a risk factor for late mortality in multivariate analysis. We were not able to demonstrate that this was related only to the IVR-ASO procedure.

Reoperations involving the right heart obviously were more frequent in patients who had a RVOT reconstruction at initial repair. In this IVR-RVOT group, 60% (28/47) of late reoperations were related to RVOT anomalies with half of reoperations related to replacement of a conduit.²⁴ Residual VSD closure was required in 10% of the cases with no difference in the 3 strategies.

The initial strategy is determined by the underlying anatomy of the DORV. The VSD location is the major determinant to choose between the available options. Decisional algorithms have been designed to choose the optimal surgical procedure according to different anatomic factors.⁸ When the initial repair is performed, late outcomes are similar in the different anatomic subtypes of DORV. This is certainly reassuring for the counseling of families, particularly when a prenatal diagnosis of DORV has been made.

Study Limitations

The main limitation of this study is in the anatomic classification of DORV, which is insufficiently complete to allow the anatomic groups to coincide with surgical strategies. The relationship of the VSD to the great arteries and the presence or absence of subpulmonary obstruction are necessary but not sufficient elements to decide on the proper surgical strategy for a DORV. This is the reason why various surgical strategies are presented for each anatomic group in our study.

Our surgical classification for 3 main groups has forced us to assemble different strategies for IVR-RVOT enlargement, such as the Lecompte or Rastelli procedures. We were unable to analyze separately the different substrategies in the IVR-RVOT group. For patients with the Taussig-Bing type undergoing a Lecompte or Rastelli procedure, we classified them in the IVR-RVOT surgical group because the aorta was not mobilized during surgery (no arterial switch by definition). That is why in our series the Taussig-Bing type does not automatically imply a classification for the IVR-ASO strategy. Risk factor analysis is by nature a predictor of outcome, which may, in turn, be influenced by varying treatment options.

CONCLUSIONS

DORV is a complex and heterogeneous anatomic group of congenital heart anomalies. Despite constant improvements in outcomes, mortality and need for reoperation remain significant because of the clinical challenge imposed by the “most complex” anatomic forms. The ncVSD type carries a higher risk of events compared with the other anatomic types of DORV, and the biventricular (vs univentricular) strategy in this setting needs to be evaluated by specific studies. The IVR-ASO

group requiring a neonatal procedure has the highest risk of early death. One third of the patients had to undergo reoperation, and the type of reoperation depends on the initial repair. The absence of difference in late mortality or reoperation among the 3 surgical strategies is certainly an important element, particularly for prenatal counseling.

Conflict of Interest Statement

Authors have nothing to disclose with regard to commercial support.

References

1. Walters HL, Mavroudis C, Tchervenkov C, Jacobs JP, Lacour-Gayet F, Jacobs ML. Congenital Heart Surgery Nomenclature and Database Project: double outlet right ventricle. *Ann Thorac Surg*. 2000;69:249-63.
2. European Association for Cardio-Thoracic Surgery, Society of Thoracic Surgeons, List S. European Paediatric Cardiac Code (IPCCC Short List). April 1, 2012. 2012:1-14. Available at: <http://ipccc.net/index.php/download-the-ipccc/>. Accessed June 13, 2016.
3. Kirklin JK, Harp RA, McGoon DC. Surgical treatment of origin of both vessels from right ventricle, including cases of pulmonary stenosis. *J Thorac Cardiovasc Surg*. 1964;48:1026-36.
4. Lecompte Y, Batisse A, Di Carlo D. Double-outlet right ventricle: a surgical synthesis. *Adv Card Surg*. 1993;4:109-36.
5. Kleinert S, Sano T, Weintraub RG, Mee RB, Karl TR, Wilkinson JL. Anatomic features and surgical strategies in double-outlet right ventricle. *Circulation*. 1997;96:1233-9.
6. Belli E, Serraf A, Lacour-Gayet F, Prodan S, Piot D, Losay J, et al. Biventricular repair for double-outlet right ventricle. Results and long-term follow-up. *Circulation*. 1998;98(19 Suppl):II360-7.
7. Artrip JH, Sauer H, Campbell DN, Mitchell MB, Haun C, Almodovar MC, et al. Biventricular repair in double outlet right ventricle: surgical results based on the STS-EACTS International Nomenclature classification. *Eur J Cardiothorac Surg*. 2006;29:545-50.
8. Li S, Ma K, Hu S, Hua Z, Yang K, Yan J, et al. Surgical outcomes of 380 patients with double outlet right ventricle who underwent biventricular repair. *J Thorac Cardiovasc Surg*. 2014;148:817-24.
9. Tchervenkov C, Walters HL, Chu VF. Congenital Heart Surgery Nomenclature and Database Project: double outlet left ventricle. *Ann Thorac Surg*. 2000;69(4 Suppl):S264-9.
10. Van Praagh R. What is the Taussig-Bing malformation? *Circulation*. 1968;38:445-9.
11. Lev M, Bharati S, Meng CC, Liberthson RR, Paul MH, Idriss F. A concept of double-outlet right ventricle. *J Thorac Cardiovasc Surg*. 1972;64:271-81.
12. Mostefa-Kara M, Bonnet D, Belli E, Fadel E, Houyel L. Anatomy of the ventricular septal defect in outflow tract defects: similarities and differences. *J Thorac Cardiovasc Surg*. 2014;149:682-8.
13. Hayes DA, Jones S, Quaegebeur JM, Richmond ME, Andrews HF, Glickstein JS, et al. Primary arterial switch operation as a strategy for total correction of Taussig-Bing anomaly: a 21-year experience. *Circulation*. 2013;128(11 Suppl 1):S194-8.
14. Belli E, Serraf A, Lacour-Gayet F, Hubler M, Zoghby J, Houyel L, et al. Double-outlet right ventricle with non-committed ventricular septal defect. *Eur J Cardiothorac Surg*. 1999;15:747-52.
15. Lacour-Gayet F, Haun C, Ntalakoura K, Belli E, Houyel L, Marcsek P, et al. Biventricular repair of double outlet right ventricle with non-committed ventricular septal defect (VSD) by VSD rerouting to the pulmonary artery and arterial switch. *Eur J Cardiothorac Surg*. 2002;21:1042-8.
16. Brown JW, Ruzmetov M, Okada Y, Vijay P, Turrentine MW. Surgical results in patients with double outlet right ventricle: a 20-year experience. *Ann Thorac Surg*. 2001;72:1630-5.
17. Patel JK, Glatz AC, Ghosh RM, Jones SM, Natarajan S, Ravishanker C, et al. Intramural ventricular septal defect is a distinct clinical entity associated with postoperative morbidity in children after repair of conotruncal anomalies. *Circulation*. 2015;132:1387-94.

18. Curzon CL, Milford-Beland S, Li JS, O'Brien SM, Jacobs JP, Jacobs ML. Cardiac surgery in infants with low birth weight is associated with increased mortality: analysis of the Society of Thoracic Surgeons Congenital Heart Database. *J Thorac Cardiovasc Surg.* 2008;135:546-51.
19. Jenkins KJ, Newburger JW, Lock JE, Davis RB, Coffman GA, Lezzoni LI. In-hospital mortality for surgical repair of congenital heart defects: preliminary observations of variation by hospital caseload. *Pediatrics.* 1995;95:323-30.
20. Prêtre R, Tamisier D, Bonhoeffer P, Mauriat P, Pouard P, Sidi D. Results of the arterial switch operation in neonates with transposed great arteries. *Lancet.* 2001;357:1826-30.
21. Vergnat M, Baruteau AE, Houyel L, Ly M, Roussin R, Capderou A, et al. Late outcomes after arterial switch operation for Taussig-Bing anomaly. *J Thorac Cardiovasc Surg.* 2015;149:1124-30.
22. Losay J, Touchot A, Serraf A, Litvinova A, Lambert V, Piot JD, et al. Late outcome after arterial switch operation for transposition of the great arteries. *Circulation.* 2001;104(Suppl 1):I-121-6.
23. Belli E, Serraf A, Lacour-Gayet F, Inamo J, Houyel L, Bruniaux J, et al. Surgical treatment of subaortic stenosis after biventricular repair of double-outlet right ventricle. *J Thorac Cardiovasc Surg.* 1996;112:1570-80.
24. Kreutzer C, De Vive J, Oppido G, Kreutzer J, Gauvreau K, Freed M, et al. Twenty-five-year experience with Rastelli repair for transposition of the great arteries. *J Thorac Cardiovasc Surg.* 2000;120:211-23.

Key Words: congenital heart disease, double-outlet right ventricle, outcome, pediatrics, surgery

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TABLE E1. List of variables considered for multivariable analyses in overall analysis

Late reoperation	Late mortality
Restrictive VSD	Restrictive VSD
Subaortic obstruction	Mitral cleft
Concomitant surgical procedure	Coronary artery anomalies
Duration of CPB	
Days of intubation	

VSD, Ventricular septal defect; CPB, cardiopulmonary bypass.

TABLE E2. Univariate analysis for late mortality

	Mortality					
	IVR (n = 149)		IVR-RVOT (n = 163)		IVR-ASO (n = 121)	
	95% CI	P	95% CI	P	95% CI	P
Demographic						
Gender	0.4-2.8	.98	0.1-1.3	.12	0.4-1.9	.69
Center	0.5-3.8	.53	0.2-2.3	.63	0.9-4.6	.08
Postnatal diagnosis	0.3-2.6	.90	0.1-1.2	.10	0.4-2.1	.88
Genetic abnormality	0.2-4.5	.64	0.4-2.9	.35	0.6-5.7	.74
Anatomy						
Fallot type	0.5-1.6	.54	0.3-1.4	.64	NA	NA
VSD type	0.3-1.8	.78	0.1-2.1	.96	NA	NA
Taussig-Bing type	NA	NA	0.2-5.2	.89	0.5-1.8	.31
ncVSD	1.2-2.2	.04	0.6-1.4	.16	1.3-2.7	.03
Multiple VSD	0.4-2.3	.36	0.1-6.9	.91	0.7-4.6	.18
Restrictive VSD	0.4-2.0	.06	0.4-9.1	.36	0.7-7.9	.16
Subpulmonary obstruction	NA	NA	0.3-2.1	.15	0.3-2.7	.83
Subaortic obstruction*	0.7-7.6	.09	2.2-15	<.01	0.7-3.6	.23
AVSD	2.4-89	<.01	0.1-9.4	.49	0.1-6.7	.30
Mitral cleft	1.3-2.1	.04	0.4-2.3	.29	1.2-31	.03
Straddling	0.1-4.3	.59	0.1-8.6	.91	0.5-2.5	.84
Mitral valve abnormality	1.3-11	<.01	0.7-1.5	.11	0.9-7.9	.06
Coronary artery anomalies	1.2-5.9	.04	0.9-2.6	.07	0.9-4.4	.15
Previous palliative surgery						
Palliative procedure	0.3-2.8	.88	0.8-8.6	.12	0.4-2.6	.95
Surgical strategy						
Date (>/<2002)	0.2-5.4	.34	0.3-2.1	.69	0.7-2.9	.24
Age	0.9-1.0	.78	0.9-1.0	.34	0.9-1.0	.23
Weight	1.0-1.1	.03	0.9-1.0	.056	0.9-1.0	.01
Concomitant procedure	2.2-18	<.01	0.2-3.5	.75	1.7-25	<.01
Duration of CPB	0.9-1.3	.11	1.0-1.1	<.01	1.0-1.1	<.01
Postsurgery						
Days of intubation	0.7-1.4	.88	1.1-1.2	.02	1.0-1.1	<.01
Early reoperation (<30 d)	0.2-1.3	.61	1.9-26	<.01	2.2-14	<.01
Late reoperation (>30 d)	0.9-8.3	.06	0.3-9.3	.61	0.1-1.5	.12
No. of reoperations >1	0.5-7.1	.41	0.7-1.3	.12	0.5-4.8	.40

IVR, Intraventricular repair; RVOT, right ventricular outflow tract; ASO, arterial switch operation; CI, confidence interval; NA, not applicable; VSD, ventricular septal defect; ncVSD, noncommitted ventricular septal defect; AVSD, atrioventricular septal defect; CPB, cardiopulmonary bypass. *Subaortic obstruction (including the patients with coarctation of aorta, interrupted aortic arch, valvular, or subvalvular aortic stenosis).

TABLE E3. Univariate analysis of overall cohort for late outcomes

	Reoperation			Mortality		
	HR	95% CI	P	HR	95% CI	P
Demographic						
Gender	1.16	0.80-1.68	.4	0.79	0.46-1.35	.4
Center	0.92	0.53-1.60	.6	1.12	0.66-1.93	.6
Postnatal diagnosis	0.96	0.66-1.41	.8	0.84	0.49-1.45	.5
Genetic abnormality			.8			.8
Anatomy						
Fallot type	0.84	0.56-1.27	.4	0.65	0.34-1.23	.2
VSD type	2.02	0.93-4.38	.1	0.88	0.38-2.04	.9
Taussig–Bing type	1.37	0.88-2.14	.1	1.92	0.97-3.82	.06
ncVSD	2.02	0.93-4.38	.08	2.99	1.06-8.46	.02
Multiple VSD	0.86	0.46-1.59	.8	.2711	0.57-2.81	.6
Restrictive VSD	1.75	0.98-3.12	.06	2.29	1.08-4.84	.03
Subpulmonary obstruction			.3			.3
Subaortic obstruction*			<.01			<.01
AVSD			.1			.06
Mitral cleft	2.06	0.76-5.57	.1	3.30	1.70-6.4100	<.01
Straddling	1.14	0.70-1.86	.6	1.28	0.64-2.53	.5
Mitral valve abnormality	1.72	0.92-3.20	.08	1.45	0.71-2.19	.4
Coronary artery anomalies	1.28	0.87-1.88	.2	1.93	1.11-3.35	.01
Previous palliative surgery						
Palliative procedure	1.17	0.80-1.70	.4	.042	0.59-1.86	.9
Surgical strategy						
Date (>/<2002)	1.34	0.85-2.09	.8	1.42	0.73-2.29	.4
Age	1.00	0.99-1.00	.1	0.99	0.99-1.00	.08
Weight	1.00	0.99-1.00	.2	0.99	0.99-1.00	.03
IVR	0.79	0.53-1.20	.3	0.82	0.45-1.48	.5
IVR-RVOT	1.08	0.68-1.72	.7	0.70	0.33-1.50	.3
IVR-ASO	1.49	0.94-2.36	.08	1.90	1.01-3.59	.04
Concomitant procedure	1.84	1.25-2.72	<.01	2.58	1.50-4.44	<.01
Duration of CPB	1.01	1.00-1.01	<.01	1.01	1.01-1.02	<.01
Postsurgery						
Days of intubation	1.05	1.04-1.07	<.01	1.04	1.02-1.07	<.01
Early reoperation (<30 d)		NA		5.06	2.51-10.19	<.01
Late reoperation (>30 d)		NA		1.04	0.40-2.69	.9
No. of reoperations >1		NA		2.57	0.52-12.62	.2

HR, Hazard ratio; CI, confidence interval; VSD, ventricular septal defect; ncVSD, noncommitted ventricular septal defect; AVSD, atrioventricular septal defect; IVR, intraventricular repair; RVOT, right ventricular outflow tract; ASO, arterial switch operation; CPB, cardiopulmonary bypass; NA, not applicable. *Subaortic obstruction (including the patients with coarctation of aorta, interrupted aortic arch, valvular, or subvalvular aortic stenosis).

TABLE E4. Multivariate analysis results of overall cohort for late outcomes (with the number of events)

Late mortality (n = 20)						
Covariate (events)	b	SE	Wald	P	Exp(b)	95% CI of Exp(b)
Restrictive VSD (n = 11)	0.9619	0.4088	5.5367	.0144	2.7113	1.2161-6.3494
Mitral cleft (n = 8)	1.3328	0.3534	14.220	.0037	3.5022	1.6193-5.5812
Coronaries artery anomalies (n = 12)	1.1243	0.6852	9.2564	.0139	2.8831	1.5016-3.922

Late reoperation (n = 97)						
Covariate	b	SE	Wald	P	Exp(b)	95% CI of Exp(b)
Concomitant surgical procedure (n = 29)	0.4917	0.2287	4.6246	.0315	1.6351	1.0469-2.5539
Duration of CPB	0.0069	0.0014	24.459	<.001	1.0070	1.0042-1.0097

SE, Standard error; CI, confidence interval; VSD, ventricular septal defect; CPB, cardiopulmonary bypass.

TABLE E5. Univariate analysis for late reoperation

	Reoperation					
	IVR (n = 149)		IVR-RVOT (n = 163)		IVR-ASO (n = 121)	
	95% CI	P	95% CI	P	95% CI	P
Demographic						
Gender	0.6-2.6	.55	0.5-1.9	.88	0.6-2.4	.48
Center	0.3-1.2	.15	0.4-1.3	.17	0.6-2.3	.52
Postnatal diagnosis	0.5-2.5	.73	0.4-1.4	.40	0.6-2.1	.73
Genetic abnormality	0.4-1.6	.59	0.8-1.7	.24	0.7-1.6	.29
Anatomy						
Fallot type	0.6-2.1	.41	0.4-2.1	.62	NA	NA
VSD type	0.2-1.8	.57	0.3-2.7	.88	NA	NA
Taussig-Bing type	NA	NA	0.4-2.4	.90	0.7-2.4	.72
ncVSD	0.4-1.1	.40	2.2-13	<.01	0.1-3.9	.54
Multiple VSD	0.1-66	.97	0.3-2.3	.69	0.3-1.9	.65
Restrictive VSD	0.1-0.9	.04	0.5-3.3	.57	1.1-6.8	.04
Subpulmonary obstruction	0.8-1.9	.16	0.4-2	.83	0.8-2.4	.24
Subaortic obstruction*	1.9-4.5	.02	0.3-1.9	.54	1.5-5.1	<.01
AVSD	2-7.1	.01	0.1-17	.95	0.1-15	<.01
Mitral cleft	0.4-5.9	.18	0.6-2.5	.16	0.9-1.9	.09
Straddling	0.3-3.1	.94	0.4-2.6	.43	0.3-1.9	.67
Mitral valve abnormality	0.2-4.3	.21	0.8-3.4	.11	0.8-4.7	.09
Coronary artery anomalies	0.8-2.6	.10	0.2-1.7	.41	0.6-1.9	.30
Previous palliative surgery						
Palliative procedure	0.7-3.0	.30	0.6-2.2	.59	0.6-2.4	.64
Surgical strategy						
Date (>/<2002)	0.6-1.8	.63	0.4-1.6	.91	0.8-2.2	.21
Age	0.9-1.0	.59	0.9-1.0	.25	0.9-1.0	.36
Weight	0.9-1.0	.86	0.9-1.0	.79	0.9-1.0	.40
Concomitant procedure	0.8-4.1	.11	0.6-2.5	.65	1.2-4.1	<.01
Duration of CPB	0.9-1.0	.052	1.0-1.1	<.01	1-1.1	<.01
Postsurgery						
Days of intubation	1-1.2	.04	1.0-1.1	<.01	1-1.1	<.01
Early reoperation (<30 d)				NA		
Late reoperation (>30 d)				NA		
No. of reoperations >1				NA		

IVR, Intraventricular repair; RVOT, right ventricular outflow tract; ASO, arterial switch operation; CI, confidence interval; NA, not applicable; VSD, ventricular septal defect; ncVSD, noncommitted ventricular septal defect; AVSD, atrioventricular septal defect; CPB, cardiopulmonary bypass. *Subaortic obstruction (including the patients with coarctation of aorta, interrupted aortic arch, valvular, or subvalvular aortic stenosis).