Echocardiographic Classification and Surgical Approaches to Double-Outlet Right Ventricle for Great Arteries Arising Almost Exclusively from the Right Ventricle

Selecting an appropriate surgical approach for double-outlet right ventricle (DORV), a complex congenital cardiac malformation with many anatomic variations, is difficult. Therefore, we determined the feasibility of using an echocardiographic classification system, which describes the anatomic variations in more precise terms than the current system does, to determine whether it could help direct surgical plans. Our system includes 8 DORV subtypes, categorized according to 3 factors: the relative positions of the great arteries (normal or abnormal), the relationship between the great arteries and the ventricular septal defect (committed or noncommitted), and the presence or absence of right ventricular outflow tract obstruction (RVOTO). Surgical approaches in 407 patients were based on their DORV subtype, as determined by echocardiography. We found that the optimal surgical management of patients classified as normal/committed/no RVOTO, normal/committed/RVOTO, and abnormal/committed/no RVOTO was, respectively, like that for patients with large ventricular septal defects, tetralogy of Fallot, and transposition of the great arteries without RVOTO. Patients with abnormal/committed/RVOTO anatomy and those with abnormal/noncommitted/RVOTO anatomy underwent intraventricular repair and double-root translocation. For patients with other types of DORV, choosing the appropriate surgical approach and biventricular repair techniques was more complex. We think that our classification system accurately groups DORV patients and enables surgeons to select the best approach for each patient’s cardiac anatomy. (Tex Heart Inst J 2017;44(4):245-51)
which describes the anatomic variations seen in DORV patients in more precise terms and provides specific surgical approaches for 8 DORV subtypes.

**Patients and Methods**

From February 2001 through July 2014, 500 patients diagnosed with DORV underwent treatment at the Fuwai Hospital. Diagnosis was based on 2-dimensional echocardiographic evidence. Cardiac catheterization and CT angiography were performed when necessary. Ultimately, 407 (81.4%) patients underwent surgery. The remaining 93 patients were not operated on because of late presentation, family choices, or a very high or prohibitive surgical risk. The study was approved by the institutional review board of Fuwai Hospital, and consent was obtained from the patients or from their parents or guardians.

We retrospectively classified the 500 patients into subgroups according to the relative positions of the great arteries (normal or abnormal), the relationship between the great arteries and the VSD (committed or noncommitted), and the presence or absence of RVOTO (Table I). Figures 1 through 4 illustrate the important anatomic characteristics that are associated with the 8 DORV subtypes and are identifiable through echocardiography (not all subtypes are shown). We then evaluated the modified Fuwai Hospital echocardiographic categorization by using the information obtained through cardiac catheterization, CT angiography, and surgical inspection; monitored the surgical outcomes of patients in the different subgroups; and clarified our surgical protocols on the basis of the detailed echocardiographic data collected.

**Diagnosis**

The diagnosis of DORV followed the 90% rule: that is, we diagnosed patients with DORV if 100% of one great artery and 90% of the others arose from the right ventricle (RV). We defined Taussig-Bing–type DORV as DORV in which 50% of the pulmonary artery (PA) overrode the VSD.

**TABLE I. Modified Fuwai Classification in 500 Patients with Double-Outlet Right Ventricle**

| DORV Subtype | No. Patients (%) | Relative Position of Great Arteries | Relationship between VSD and Great Arteries | RVOTO |
|--------------|------------------|------------------------------------|------------------------------------------|-------|
| I-A          | 89 (17.8)        | Normal                             | Committed                                | Absent|
| I-B          | 89 (17.8)        | Normal                             | Committed                                | Present|
| II-A         | 33 (6.6)         | Normal                             | Noncommitted                             | Absent|
| II-B         | 32 (6.4)         | Normal                             | Noncommitted                             | Present|
| III-A        | 62 (12.4)        | Abnormal                           | Committed                                | Absent|
| III-B        | 60 (12.0)        | Abnormal                           | Committed                                | Present|
| IV-A         | 31 (6.2)         | Abnormal                           | Noncommitted                             | Absent|
| IV-B         | 104 (20.8)       | Abnormal                           | Noncommitted                             | Present|

DORV = double-outlet right ventricle; RVOTO = right ventricular outflow tract obstruction; VSD = ventricular septal defect

**Anatomic Features of DORV.** The VSD in DORV was described as committed or noncommitted according to the distance from the superior margin of the VSD to

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**Fig. 1** Transthoracic echocardiograms (parasternal long-axis views) of a 10-month-old boy with type I-B double-outlet right ventricle show A) a large ventricular septal defect (asterisk) under the aorta and the aorta overriding the defect, and B) an aortic trunk located posterior to and to the right of the hypoplastic pulmonary trunk, that is, a normal positional relationship of the great arteries.

Ao = aorta; LPA = left pulmonary artery; MPA = main pulmonary artery; RPA = right pulmonary artery.
Fig. 2 Transthoracic echocardiograms (parasternal long-axis views) in a 2-month-old girl with type II-A double-outlet right ventricle show A) a noncommitted ventricular septal defect (asterisk) far from the arterial valves, and abnormal mitral–aortic fibrous continuity, and B) 2 great arteries spiraling around each other in a normal pattern, without pulmonary outflow tract obstruction.

Aq = aorta; PA = pulmonary artery

Fig. 3 Transthoracic echocardiograms (parasternal long-axis views) in a 4-month-old girl with type III-A double-outlet right ventricle show A) a subpulmonary ventricular septal defect (asterisk), and B) an aortic trunk anterior to the dilated pulmonary trunk.

Aq = aorta; PA = pulmonary artery

Fig. 4 Transthoracic echocardiograms (parasternal long-axis views) in a one-year-old boy with type IV-A double-outlet right ventricle show A) a noncommitted ventricular septal defect (asterisk) very remote from the great arteries and with long, muscular continuity between the mitral and aortic valves, and B) a ventricular septal defect (asterisk) in the inlet septum.

Aq = aorta
the center of the arterial valves. If the distance was less than or equal to the aortic diameter, the VSD was committed; if the distance was greater than the aortic diameter, the VSD was noncommitted and was typically located in the inlet septum (sometimes with perimembranous extension), the trabecular septum, or both. An RVOTO can be infundibular or purely valvular, with or without a small pulmonary valve annulus and hypoplasia of the PAs. An RVOTO was diagnosed if the patient had a peak systolic gradient >30 mmHg.

Statistical Analysis
Analyses were performed by using SPSS version 18 (IBM Corporation; Endicott, NY). We used the Student t test to analyze continuous variables, and we used the Fisher exact test to compare discrete variables. A P value <0.05 was considered statistically significant.

Results
Of the 500 patients, there were 299 males and 201 females. The mean age at diagnosis was 4.08 years (range, 6d-42 yr), and the mean weight was 13.3 ± 11.39 kg (range, 2.1–94 kg). The patients’ oxygen saturation levels ranged from 25% to 94% without supplemental oxygen (Table II).

Of the 500 patients, 407 (81.4%) underwent surgery. Of those, 272 (66.8%) had biventricular repair, 99 (24.3%) had single-ventricle repair, and 36 (8.8%) had palliative procedures. At the time of surgery, the patients’ ages ranged from 4 months to 42 years, and 208 (51.1%) were younger than 2 years old. Cardiac catheterization and CT angiography were performed in 264 (52.8%) and 59 (11.8%) of the 500 patients, respectively. When we compared the findings of cardiac catheterization, CT angiography, and intraoperative inspection with the diagnosis made through preoperative echocardiography, we found agreement regarding the relative position of the great arteries in 414 of 414 patients (accuracy rate, 100%), and we found agreement regarding the relationship between the great arteries and the VSD in 347 of 378 patients (accuracy rate, 91.8%).

Table III shows the surgical management protocols for each subtype. The surgical success rate was 92.1% for the DORV operations performed on patients categorized according to the modified Fuwai echocardiographic classification system. Of note, the surgical treatment of patients with DORV subtypes I-A, I-B, and III-A was similar.

In 22 patients with type II-A DORV, biventricular repairs were made; this involved intraventricular tunnel repair (IVR) from the left ventricle to the aorta and some additional techniques as needed, including enlargement of the VSD, use of multiple patches on the tunnel, resection of muscular bands, or reattachment of the tricuspid valve’s chordae tendineae. An IVR from the left ventricle to the aorta was performed in 11 patients with type II-B, in 14 with type III-B, and in 18 with type IV-B DORV; these patients also underwent concomitant reconstruction of the RV outflow tract performed by means of a reparation à l’etage ventriculaire (REV) procedure, Rastelli procedure, or RV–PA conduit. The tricuspid papillary muscle was reimplanted on the tunnel patch in 2 patients with type II-A DORV, in one with type II-B, and in one with type IV-B.

### Table II. Clinical Characteristics of Patients with Different Types of DORV

| Variable   | I-A (n=89) | I-B (n=33) | II-A (n=32) | II-B (n=62) | III-A (n=60) | III-B (n=31) | IV-A (n=104) | IV-B (n=30) |
|------------|------------|------------|-------------|-------------|-------------|-------------|--------------|-------------|
| Age at diagnosis (yr) | 3.72 ± 6.33 | 4.07 ± 6.26 | 4.71 ± 5.84 | 4.52 ± 7.69 | 2.78 ± 5.04 | 4.72 ± 5.08 | 2.60 ± 2.56 | 4.94 ± 5.59 |
| (2 mo–30 yr) | (1.4 mo–34 yr) | (1.3 mo–30 yr) | (4 mo–42 yr) | (0.3 mo–26 yr) | (0.8 mo–22 yr) | (1.2 mo–10 yr) | (0.2 mo–31 yr) |
| Height (cm) | 79.75 ± 26.82 | 88.3 ± 26.21 | 87.11 ± 27.87 | 96.13 ± 27.58 | 74.43 ± 29.84 | 92.82 ± 25.6 | 79.19 ± 27.62 | 96.22 ± 28.2 |
| Weight (kg) | 10.69 ± 8.68 | 13.46 ± 10.83 | 12.12 ± 9.57 | 16.31 ± 13.41 | 10.44 ± 11.82 | 14.41 ± 9.17 | 12.97 ± 13.68 | 16.32 ± 13.2 |
| SBP (mmHg) | 93.3 ± 11.4 | 97.75 ± 11.89 | 94.46 ± 11.61 | 98.72 ± 11.89 | 91.72 ± 18.16 | 97.51 ± 12.89 | 92.5 ± 12.59 | 102.9 ± 15.92 |
| DBP (mmHg) | 56.13 ± 9.32 | 60.37 ± 12.28 | 55.96 ± 12.97 | 60.47 ± 11.76 | 54.9 ± 11.79 | 60.16 ± 12.1 | 58.12 ± 12.46 | 63.57 ± 11.91 |
| SPO2 (%) | 91.75 ± 7 | 82.29 ± 11.15 | 88.69 ± 5.46 | 78.52 ± 9.33 | 78.39 ± 11.12 | 75.6 ± 12.02 | 83.81 ± 8.84 | 76.69 ± 12.24 |

SBP = systolic blood pressure; DORV = double-outlet right ventricle; DBP = diastolic blood pressure; SPO2 = arterial blood oxygen saturation

Data are presented as mean (range) or as mean ± SD.
Double-root translocation with baffling of the VSD to the neo-aorta was performed in 5 patients with type III-B and in 6 patients with type IV-B DORV. An arterial switch operation (ASO) was performed after successful relief of pulmonary valvular and subpulmonary stenosis in 4 patients with type III-B and in 6 patients with type IV-A DORV. Atrioventricular discord was present in 2 patients with type III-B and in one patient with type IV-B DORV; they underwent a Mustard or Senning atrial baffle procedure.9,10

Follow-up information was available for 396 patients (mean, 1.1 yr; range, 0.5 mo–10.8 yr). During this time, 10 patients underwent surgery to relieve subaortic or subpulmonary stenosis and to resect an RV outflow tract aneurysm.11,12

Discussion

Our study had 3 principal findings: 1) the modified Fuwai echocardiographic classification system, which is based on the relative positions of the great arteries, the relationships of arteries and the VSD, and the presence or absence of a RVOTO, is valid and feasible; 2) every subtype of DORV needs different, specific surgical approaches, a finding confirmed by clinical observations; and 3) the success rate for DORV operations performed on patients who had been categorized according to the modified Fuwai echocardiographic classification system was 92.1%.

Because our aim was to present the typical echocardiographic characteristics needed to classify DORV, we used a strict definition—the 90% rule—to select patients with DORV, and we were especially careful to distinguish TOF-type DORV (type I-B) from TOF. In terms of surgical repair, type I-B DORV is technically much more challenging than TOF is.

For patients with type I-A DORV who had aortic–mitral muscular separation or long fibrous continuity, the optimal surgical approach was an IVR from the left ventricle to the aorta; if a restrictive VSD was present, it was enlarged superiorly or anteriorly. Patients with type I-B DORV had a significant anterior malposition of the aorta that was repaired by using techniques similar to those described for the repair of TOF.

Type II DORV was characterized by normal spatial relationships of the great arteries and a remote VSD, and was repaired with a long tunnel connecting the left ventricle to the aorta. In patients with type II DORV, the width of the tunnel should be larger than that of the aortic annulus, and the tunnel should avoid the tricuspid inflow chamber as determined echocardiographically. In some cases, a tunnel necessitated

**TABLE III. Surgical Protocols and Reasons for Reoperation by Type of DORV**

| DORV Type | Biventricular Repair (n) | Staged Palliation | Single-Ventricle Repair | Reasons for Reoperation (n) |
|-----------|--------------------------|-------------------|-------------------------|-----------------------------|
| I-A       | IVR–Ao (72)              | PA banding (3)    | —                       | LVOTO (2); RVOTO (1)        |
| I-B       | IVR–Ao + RVOT reconstruction (65) | Ao–PA shunt (7) | Fontan (7) | RVOTO (4) | RVOT aneurysm (1) |
| II-A      | IVR–Ao (22) + VSD enlargement | PA banding (4) | —                       | LVOT (1); RVOT (1)         |
| II-B      | IVR–Ao (10) + relief of RVOTO IVR–Ao + RV–PA conduit (1) | Ao–PA shunt (1) | Fontan (20) | AV valve repair (1)b |
| III-A     | IVR + ASO (45)           | PA banding (1)    | —                       | RVOT (1); TVP (1)          |
| III-B     | IVR + ASO (4)            | Ao–PA shunt (6)   | Fontan (22)             | RVOTO (2)                  |
|           | IVR + DRT (5)            |                   | —                       | —                          |
|           | IVR–Ao (14) + relief of RVOTO, Rastelli, REV, or RV–PA conduit | — | — | — |
| IV-A      | IVR + ASO (6)            | PA banding (11)   | —                       | LVOTO (1)                  |
| IV-B      | IVR + DRT (6)            | Ao–PA shunt (3)   | Fontan (50)             | LVOTO (1) |
|           | IVR–Ao (18) + relief of RVOTO, Rastelli, REV, or RV–PA conduit | — | — | RVOTO (2) |

Ao = aorta; ASO = arterial switch operation; AV = atrioventricular; DORV = double-outlet right ventricle; DRT = double-root translocation; IVR = intraventricular tunnel repair; LVOTO = left ventricular outflow tract obstruction; PA = pulmonary artery; REV = reparatio à l’étage ventriculaire; RV = right ventricle; RVOT = right ventricular outflow tract; RVOTO = right ventricular outflow tract obstruction; TVP = tricuspid valve repair; VSD = ventricular septal defect

a Some patients underwent reoperation for more than one reason.

b This patient had an atrioventricular septal defect. A second atrioventricular valvular repair was made after the first concomitant repair failed.

c Pulmonary valves were used and subpulmonary stenosis was relieved easily.
reimplantation of the tricuspid papillary muscle on the tunnel patch, as well as VSD enlargement. Evaluating the requirements for tunnel creation was the most important aspect of using echocardiograms for surgical planning. In our patients with type II-A DORV, the very remote VSDs were always in the inlet septum without extension to the perimembranous region; thus, anatomic repair was ill-advised. In these circumstances, PA banding should be attempted first in a young child or infant, because this enables single-ventricle repair in the future. In both type II-A and type II-B patients, IVR-to-aorta techniques can be performed, with additional relief of RVOTO. In type II DORV—as in type I DORV—the relationship of the great arteries is normal, so no ASO procedures are involved.

Because type III and type IV DORV are characterized by abnormal positions of the great arteries, biventricular repair might necessitate reestablishing the normal spatial relations of the great arteries. Most patients with type III-A DORV have Taussig-Bing malformations, with 50% to 100% of the PA overriding the VSD and no RVOTO. These patients are always treated with an IVR and an ASO. Type III-B DORV is characterized by the presence of RVOTO, as well as a PA overriding the VSD, which could be easily confused with a TGA in association with pulmonary stenosis. In our opinion, the diagnosis of type III-B DORV should be made only in patients with a muscular or long pulmonary–mitral fibrous connection and a ≥90% overriding of the PA. Because subaortic coni are always present in these patients, a double-root translocation is the optimal treatment. If preoperative echocardiograms show the 2 semilunar valvular annuli without well-developed, subaortic muscular coni, an IVR to the aorta—in conjunction with other procedures, including VSD enlargement, muscular band resection and REV, Rastelli procedures, or an RV–PA conduit—might be indicated instead.

Type IV-A DORV is similar to type III-A in that it is characterized by TGA; however, it also has a remote VSD in which both great arteries arise from the RV with bilateral coni. In our type IV-A patients, we performed a biventricular repair to connect the left ventricle with the PA, and we then performed an ASO. Of note, the palliative repairs and the therapeutic regimens that we used for patients with type II-A and IV-A DORV were similar.

Type IV-B DORV—the most complex subset of DORV in our cohort—was similar to type IV-A with RVOTO and needed 1) the use of multiple patches, resection of muscular bands, and reattachment of the tricuspid valve’s chordae tendinae; or 2) the reestablishment of RV-to-PA continuity by means of REV or Rastelli techniques. As is the case in patients with type II-A and IV-A DORV, echocardiograms can help to establish the size of the tunnel. If patients with type IV-B DORV have well-developed subaortic coni, a double-root translocation with tunneling of the VSD to the PA is preferable.

In our cohort, patients with associated atrioventricular septal defects (AVSDs) had mainly Rastelli type-C AVSDs, which can occur in every type of DORV except type III-A. The VSD in these patients is usually in the inlet septum, far from both arteries, and is further separated from the great arteries by the tricuspid inflow chamber. These very remote VSDs cannot be connected to the great arteries unless a large superior component extends close to the aortic valve annulus. In our study, 10 of 51 patients with an AVSD underwent complete biventricular repairs despite the complex techniques necessary during the surgery.

Conclusions

In our large patient series, the modified Fuwai Hospital echocardiographic classification system for DORV accurately grouped patients, provided surgeons with precise anatomic data, and enabled surgeons to select the best possible surgical approaches for the patients’ cardiac anatomy. The accuracy of the echocardiographic classification was verified by cardiac CT angiography, cardiac catheterization, and surgical results. We think that this integrated approach of echocardiographic imaging and preoperative planning improves the surgical management of high-risk patients.

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