Biventricular repair in double outlet right ventricle: surgical results based on the STS-EACTS International Nomenclature classification

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Abstract

Background: The STS-EACTS International Nomenclature for Congenital Heart Surgery (CHS) defines four anatomic subtypes of double outlet right ventricle (DORV) based on the relationship of the ventricular septal defect (VSD) with the great vessels and the presence of right ventricular outflow tract obstruction (RVOTO). We reviewed our experience with DORV patients and two ventricles that underwent repair, applying this nomenclature. Methods: Between January 2000 and January 2005, 50 patients with DORV and two viable ventricles underwent surgical intervention: 44 patients had biventricular repair, 3 had \( n = 1 \) ventricular repair, 2 underwent a Fontan, and 1 died prior to corrective surgery. Median age at repair was 9.1 months (range: 4D—4Y). Eighteen patients (36%) were DORV-Fallot (including 5 with AVSD and heterotaxy), 9 (18%) were DORV-TGA (Taussig-Bing), 12 (24%) were DORV-VSD, and 11 (22%) were DORV non-committed VSD. Corrective surgery included 35 repairs with a VSD-aorta baffle \( \rightarrow \) RVOTO procedure and 12 arterial switches with a VSD-PA baffle (9 Taussig-Bing and 3 DORV-ncVSD). Associated procedures included 13 VSD enlargements, 8 subaortic resections, 9 arch repairs, 5 AVSD repairs, and 7 others. Results: There were three deaths in the 50 patients studied (overall mortality of 6%). Excluding one patient that died prior to corrective surgery and the two patients palliated with a Fontan procedure, the actual surgical mortality for a corrective repair was 4.3% (2/47 patients). Two surgical deaths occurred following, respectively, one repair of a Taussig-Bing with an interrupted arch and a Swiss cheese VSD and one repair of ncVSD-type with pulmonary atresia that had undergone a previous cavo-pulmonary anastomosis. No late deaths occurred. Two late reoperations included a heart transplant in a DORV-Fallot patient with Swiss cheese VSD and subaortic resection in a DORV-ncVSD patient. Angioplasties were needed for PA stenosis (\( n = 2 \)) and aortic arch obstruction (\( n = 2 \)). Four patients had LV to aorta baffle gradients between 10 and 20 mmHg. All patients were of NYHA class I/II. Conclusions: The STS-EACTS International Nomenclature provides more uniform analysis of outcomes with respect to acceptable surgical risk and mortality. Biventricular repair can be safely achieved on selected DORV, including DORV-ncVSD and DORV with AVSD and heterotaxy lesions traditionally indicated for a single ventricle palliative approach.

Keywords: Congenital heart surgery; Double outlet right ventricle; Cyanotic CHD; Not cyanotic CHD; Malposition of the great arteries

1. Introduction

Double outlet right ventricle (DORV) is characterized by a malposition of the great arteries. It is a primitive mode of ventriculoarterial connection with the aorta arising predominantly from the right ventricle \cite{1}. DORV can be part of complex anatomy including that of a single ventricle or it can occur with two functional ventricles representing a distinct clinical entity. This latter presentation exhibits a wide spectrum of anatomic and physiologic variation. Depending on the location of the ventricular septal defect (VSD), the degree of malposition of the great vessels, and the presence of right ventricular outflow tract obstruction (RVOTO), DORV can mimic clinically an unrestrictive VSD, a tetralogy of Fallot (TOF) or a transposition of the great arteries (TGA). This heterogeneity has naturally led to controversies over the anatomical definition, classification schemes, and the techniques for and the timing of surgical repair.

Traditional classification of DORV based on the relational anatomy between the VSD and the great arteries \cite{2} provides useful anatomical information; however, there is no absolute correlation between the commitment of the VSD to the great arteries and the surgical approach \cite{3}. Recent STS-EACTS International Nomenclature adopted by the databases of the Society of Thoracic Surgeons (STS) and European Association of Cardiothoracic Surgery (EACTS) \cite{4,5} and also by the
Association for European Pediatric Cardiology (AEPCC) [6] defines four types of DORV based rather on the clinical presentation and treatment: (1) VSD-type, (2) Fallot-type, (3) TGA-type, and (4) DORV non-committed VSD. This system offers uniformity to DORV nomenclature. The STS-EACTS International Nomenclature includes a limited number of items called ‘short lists’ [4,5]. The group of DORV and AVSD has not been isolated in this diagnoses short list. Its correct categorization either in the group of DORV-ncVSD or in the group of DORV-Fallot has remained controversial. The purpose of this surgical study is to retrospectively analyze a group of 50 patients with DORV and two viable ventricles, using the STS-EACTS International Nomenclature for CHS; by focusing more precisely on the groups of DORV-ncVSD and DORV-AVSD.

2. Methods

2.1. Patients and definitions

Between January 2000 and June 2005, 50 patients with DORV, concordant atroventricular connections, and two viable ventricles underwent surgical intervention at Denver Children’s Hospital and the Eppendorf University Hospital. Diagnosis was made by standard 2D echocardiography. The term ‘double outlet’ was used to describe a ventriculoarterial connection in which more than 50% of both arterial valves are connected to the same ventricle [7]. To avoid subjectivity of the ‘50% rule’, the presence of a subaortic conus or the absence of mitro-aortic fibrous continuity [8] (or mitro-pulmonary continuity for patients with transposed great arteries) was used as secondary criterion when the diagnosis was in question. The diagnosis of DORV was confirmed by two independent echocardiographers and the patients were classified according to the four types of DORV outlined by the STS and EACTS:

1. VSD-type: DORV with subaortic VSD.
2. Fallot-type: DORV with subaortic or double committed VSD and pulmonary outflow stenosis,
3. TGA-type (Taussig-Bing): DORV with a subpulmonary VSD,
4. Non-committed VSD-type — DORV with a remote VSD.

The latter term, non-committed VSD [2] remains a matter of confusion and debate. The important relationship between the arterial valves and the VSD has been described as a ‘considerable’ distance [8] or a distance superior than the aortic diameter [9]. The VSD itself has been described as either exclusively a muscular inlet type or an AV canal type VSD [10]. A more specific definition of ncVSD-type that was used throughout our analysis defines the distance between the VSD and both the aortic and pulmonary annulus as a length greater than the aortic diameter. Additionally, both vessels arise 100% from the right ventricle and there is constantly the presence of a double conus [11,12]. Importantly, this definition excludes the Taussig-Bing heart and the DORV heart with a complete AVSD. In Taussig-Bing heart, the VSD is located above the trebecula septomarginals in close proximity to the pulmonary valve [19]. Furthermore, the pulmonary valve is not committed 100% to the RV. As for the DORV heart with a complete AVSD, the VSD component uniformly has outlet extension underneath the aorta and there is constantly an associated pulmonary valvular and infundibular stenosis. This form resembles the AV canal form of tetralogy of Fallot, and for this reason is categorized in this study with the Fallot-type DORV. Such taxonomy strays from the international nomenclature [4] where the AVSD-type DORV is not defined [4,5] in the short lists forming the international nomenclature (hierarchy levels 1 and 2) or categorized with the ncVSD-type [4] in the comprehensive lists (hierarchy levels 3, 4, 5, etc.).

3. Results

Feasibility of biventricular repair was considered in all patients based on the adequacy of both ventricles, evaluated at 2D echocardiography. The surgical procedures were performed by one of three surgeons (F.L.G., V.H., D.N.C.) and was determined along with the referring cardiologist after considering all clinical data including associated cardiac and non-cardiac malformations, previous operations, and associated genetic disorders. Table 1 illustrates the complex characteristics of the patients with DORV and two viable ventricles. The Fallot-type of DORV represented the largest fraction of patients in this series, followed by the ncVSD-type, VSD-type, and TGA-type. Twenty-two patients (44%) had significant other major cardiac malformations including pulmonary atresia, total anomalous pulmonary venous return (TAPVR), complete AV septal defect (AVSD), aortic coarctation, or interrupted IVC. The most common other major cardiac malformation occurring with the Fallot-type DORV was pulmonary atresia (n = 6) followed by AVSD (n = 5). Importantly, included in the Fallot-type DORV are five patients with visceral heterotaxy syndrome. These five patients exhibited the most complex anatomy of all patients examined. Three patients had a large right-sided liver extending across the midline, asplenia, right atrial isomerism, and infradiaphragmatic TAPVR. The two other patients had a midline liver, polysplenia, left atrial isomerism, and an interrupted IVC. All five patients had a complete AVSD with the VSD extending into the outlet septum below the aortic valve and two patients had pulmonary atresia with ductal dependant pulmonary circulations. Interestingly, none of these patients had an identified genetic syndrome. The TGA-type DORV commonly had aortic arch hypoplasia with coarctation (7/9 patients). The frequency of associated other major cardiac malformations for the ncVSD-type and VSD-type DORV was 36% and 17%, respectively.

Twenty-five patients had initial palliative procedures (Table 1). The staged approach was common with the Fallot-type and ncVSD-type of DORV with a respective 83% and 73% of patients undergoing initial palliative procedures. The most frequent procedure performed for the Fallot-type was a BT shunt and for the ncVSD-type was a PA banding. All three patients presenting with the diagnosis of TAPVR underwent a pulmonary veins repair within the first few days of life with two of these patients requiring the addition of a BT shunt. Four patients underwent cavo-pulmonary shunts in anticipation of single ventricle palliation. These procedures were performed at outside institutions prior to referral for...
corrective repair. Apart, one DORV-ncVSD patient had an associated total ectopia cordis requiring a neonatal skin flap coverage.

Of the 50 patients presenting with DORV and two viable ventricles, 44 had two-ventricle repairs, 3 had 1½ ventricle repairs, two underwent a subsequent Fontan procedure and one patient died following initial palliation. A Fontan procedure was performed in a Fallot-type DORV with multiple VSDs and Down’s syndrome in a patient that had previously undergone bilateral cavopulmonary anastamosis for bilateral SVCs. The other patient diverted to a Fontan had a ncVSD-type DORV with a straddling mitral valve and a ‘smallish’ RV. The patient that died following initial palliation was a Fallot-type DORV with severe infundibular pulmonary stenosis, a restrictive VSD, and CHARGE1 syndrome. The patient underwent a BT shunt and VSD enlargement and developed progressive heart failure and acidosis. For the remaining 47 patients, the primary repair and associated major procedures performed at the time of corrective surgery are listed in Table 2. The mean age and weight at corrective repair was 273 ± 72 days and 5.8 ± 0.6 kg, respectively, and this varied with DORV-type (Table 2). Corrective surgery included 35 repairs with a VSD-aorta baffle ± RVOT procedure and 12 arterial switches with a VSD-PA baffle. Associated procedures included 13 VSD enlargements, 8 subaortic resections, 9 arch repairs, and 5 AVSD repairs. Other procedures not listed in Table 2 include takedown (n = 2) and creation (n = 1) of cavopulmonary shunts, PA plasties (n = 4), and intraatrial baffles for LSVC draining into the left atrium (n = 1).

There were three deaths in the 50 patients studied; overall mortality of 6%. Excluding the patient that died prior to corrective surgery and the two patients that palliated with a Fontan procedure, the actual surgical mortality for a corrective repair is 4.3% (2/47 patients). One neonate died following a complex arterial switch operation and VSD-PA tunnelization for a TGA-type DORV. This child was 2.4 kg and had four VSDs (‘swiss cheese’ VSD), an interrupted aortic arch, and a severe subaortic obstruction. The other death occurred in a 4-year-old child with ncVSD-type DORV and pulmonary atresia who had undergone a previous cavopulmonary anastamosis at an outside institution. The VSD was tunneled to the aorta, an RV-PA conduit placed and the cavopulmonary anastamosis taken down. The patient required postoperative extracorporeal circulatory support for RV failure and subsequently developed mediastinitis. There was one late reoperation in a patient with ncVSD-type DORV who underwent VSD-PA baffle and an arterial switch operation. This patient had a rather long duration of PA banding prior to repair and developed obstruction in the LVOT requiring a patch enlargement and muscular resection. Two patients required permanent pacemakers, both patients were ncVSD-type and had VSD enlargements. The postoperative course was related to the severity of the DORV. In the groups of DORV-TGA, DORV-AVSD, and DORV-ncVSD, there were 14

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1 Coloboma, heart disease, atresia choanae, retarded growth and development and/or central nervous system abnormalities, genital hypoplasia, and ear anomalies and/or deafness.
There were no deaths in the 12 patients presenting with this patient who presented with a necrotizing enterocolitis. In our series, a staged approach was necessary in one patient who required ECMO. There were no late deaths; however, one patient required heart transplantation for progressive LV failure. This patient was Fallot-type DORV with a ‘Swiss cheese’ VSD. At a mean follow-up of 20 months, post-operative angioplasties were needed for PA stenosis (n = 2) and aortic arch obstruction (n = 2). Four patients had LV-aorta baffle gradients between 10 and 20 mmHg. All patients were of NYHA class I/II.

4. Discussion

Biventricular repair of DORV remains a surgical challenge; however, successful repair can be achieved with less than 5% mortality. Despite these overall favorable results, the actual risk of surgical death is more specifically related to the type of lesion encountered. Early classification schemes based on the location of the VSD in relation to the great arteries unavoidably combines simple and complex lesions within the same group despite the vastly different operative approaches. Examining outcomes according to clinical types of DORV as outlined by the new international nomenclature provides a more accurate interpretation of the data. In other words, a DORV with a simple subaortic VSD (VSD-type) more closely resembles an unrestricted VSD in respect to clinical presentation, surgical approach, and operative risk than other types of DORV. In our series, the mortality rates for the specific types of DORV were as follows: VSD-type 0%, Fallot-type 6%, ncVSD-type 9%, and TGA-type 11%. These rates are more reflective of the complexity of each type lesion and correspond more appropriately to the non-DORV sister lesion than other clinical types of DORV.

4.1. VSD-type

These patients present with clinical signs of over-circulation from an unrestricted VSD and usually require a one-stage biventricular repair within the first 6 months of life. In our series, a staged approach was necessary in one patient who presented with a necrotizing enterocolitis. There were no deaths in the 12 patients presenting with this lesion. The VSD was closed from the right atrium in four patients and through a right ventriculotomy in the remaining eight patients. Importantly, VSD enlargements were required in 42% (5/12). The frequent requirement for a VSD enlargement and a right ventricular approach distinguishes this lesion from other VSDs. These maneuvers add relatively little risk to the surgical procedure.

4.2. TGA-type (Taussig-Bing)

These patients present uniformly in the neonatal period with cyanosis typical of transposition physiology. Complete repair with an arterial switch operation and a VSD-PA baffle is required in the neonatal period. All nine of our patients were repaired in the neonatal period. Arch hypoplasia requiring reconstruction was common presenting in seven patients (78%). A subaortic muscular resection was performed in four patients during the time of corrective repair. There was one death as previously mentioned that yields a surgical mortality of 11%. This is consistent with other published results [13] for this lesion, and is comparable to the non-DORV sister lesion TGA/VSD which carries a surgical mortality rate between 5% and 10% in recent series [14,15]. Difficulties encountered with the DORV lesion likely reflect the higher frequency of complex coronary anatomy and associated subaortic obstruction.

4.3. Fallot-type

In our series, this DORV-type represents a rather heterogenous group of patients, primarily because of our inclusion of patients with heterotaxy and complete AVSDs. In our series, the VSD consistently had a large bridging anterior leaflet (Rastelli C type) found almost exclusively with TOF/AVSD. In fact, the intraventricular repair approximates the surgical repair of a TOF/AVSD, with the exception that the aorta being 100% on the RV places this repair at further risk for subaortic obstruction. Additionally, these patients have heterotaxy syndrome and frequent associated cardiac lesions such as
TAPVR or anomalous systemic venous drainage requiring alteration of the surgical strategy. The TAPVR requires repair in the neonatal period often with an accompanying BT shunt. An isolated LSVC draining into the left atrium may require a left-sided cavopulmonary anastomosis or tunneling towards the right atrium. Curiously, children with this very severe lesion of DORV-AVSD and heterotaxy do not usually have any identified genetic anomaly [16]; they are not Downs (in comparison, TOF-AVSD are ~90% Downs). Clearly, the heterotaxy group of DORV present with a higher level of surgical complexity for biventricular repair. For this reason, these children are often diverted toward a Fontan pathway [17]. In our series, we successfully repaired five patients with this lesion with zero mortality; however, long-term follow-up is lacking.

4.4. ncVSD-type

Patients with a ncVSD present with a remote VSD. Importantly, the VSD lies at a distance from both the aortic and pulmonary annulus greater than the aortic diameter [9,11]. There is complete mixing at the ventricular level, however, the VSD may be restrictive resulting in varying degrees of desaturation and over-circulation. Because of the heterogenous presentation and lack of suitable non-DORV sister lesion, we consider this the ‘true’ DORV. Depending on the easiest way to construct the intraventricular tunnel to avoid subvalvar obstruction, this lesion was treated by tunnelization to the aorta or the PA with an arterial switch operation. It was rarely directed toward a Fontan pathway. Only one Fontan procedure was performed in a patient with a straddling mitral valve type C. In our series, 7/10 patients (70%) were tunnelized to the aorta and almost all of these patients required a VSD enlargement and half required a subaortic resection of aorta and almost all of these patients required a VSD enlargement and half required a subaortic resection of aorta and almost all of these patients required a VSD enlargement and half required a subaortic resection of aorta. These maneuvers are important to prevent subvalvar obstruction [12]. Additionally, we have adopted a policy of delaying surgical repair later into infancy, a time we believe more suited for complex intraventricular repairs requiring prolonged crossclamp times. There was one death in our series of patients, 10% surgical mortality rate.

In summary, DORV is a heterogenous group of lesions that may best be classified by a clinical CHS international nomenclature suggested by STS and EACTS. This classification scheme, we believe, provides more uniform analysis of outcomes with respect to acceptable surgical risk and mortality. The lesions that remain a surgical challenge are ncVSD-type DORV and Fallot-type DORV with a complete AVSD in the setting of heterotaxy syndrome. We suggest that DORV-AVSD and heterotaxy are categorized in the group of DORV-Fallot. Nevertheless, both of these lesions have traditionally been indications for a single ventricle palliative approach [17,18]. In our series, there was one death in 15 of these challenging patients (6.7% mortality) following biventricular repair. These lesions should be considered for biventricular repair. Long-term results of biventricular repair of complex DORV are not available today. These data would be crucial in the future to compare the respective performance of biventricular repair and Fontan in complex DORV with two viable ventricles.

References


Appendix A. Conference discussion

Dr. G. Steffin (Padova, Italy): I’m concerned about the fact that you want to change the definition of double outlet right ventricle which was made by Lev and Barathi in 1972 and then revisited by others, in the sense that both vessels don’t need necessarily to be aligned to the right ventricle in order to be defined ‘double outlet right ventricle’. In that picture that you showed in your angiography, the pulmonary artery wasn’t really totally aligned to the right ventricle but only partially. I believe it is wise to maintain the old definition of double outlet right ventricle in the sense that both vessels are alligned for 50% or 60% to the right ventricle.
Dr Artrip: Absolutely. The presence of bilateral conus definitely confirms, at least in my eyes, the definition of a double outlet right ventricle.

But I’ve seen cases, actually Dr Anderson came and spoke at Denver and actually showed cases where the patients had clearly malaligned great arteries on the right ventricle without a double conus.

Dr B. Maruszewski (Warsaw, Poland): I would like to support what Giovanni just said, because with this definition, which is of course very extreme definition, 200%, you should exclude all the tetralogy of Fallot-type of double outlet right ventricle out of the series. And Francois was working on this terminology in the literature for years and actually he accepted this EACTS-STS nomenclature that we are using now.

But I would like to concentrate on the noncommitted VSD. You have a beautiful result, and congratulations on that of course, but could you allude a little bit, what was the mean age, what was the age of those patients who you repaired one-stage non-committed VSD?

Dr Artrip: Actually, the group of noncommitted VSDs, we actually don’t repair them in the neonatal period. One patient was repaired in infancy without going under a primary procedure. These kids will either get B-T shunts or get pulmonary banding, depending on which particular pathophysiology they present with.

As for approximately the age at which we did it, it was usually somewhere between 6 and 9 months. I do not recall the exact age. The other question was residual arterial gradients. We have had two patients that did have gradients about 10–20 mmHg at the end of our results. So still not insignificant, but not significant enough to require reoperation, and these are being closely followed by our cardiologists.

As for residual shunts, I don’t know the number of small residual shunts, but we had no residual shunts that were significant to require reoperation.