Chapter 24
Biventricular Repair of Double Outlet Right Ventricle with Complete Atrioventricular Septal Defect

Ming-Sing Si, Richard G. Ohye, Jennifer C. Hirsch-Romano, and Edward L. Bove

Abstract The combination of double outlet right ventricle and complete atrioventricular septal defect (DORV-AVSD) is a rare congenital heart malformation. Double outlet right ventricle with complete atrioventricular septal defect is commonly associated with heterotaxy but is distinct from DORV as associated with tetralogy of Fallot. Biventricular repair of DORV-AVSD is challenging because it requires not only correction of the complete AVSD and DORV, but often also involves resection of the outlet portion of the ventricular septum to create an unobstructed left ventricular outflow tract as well as repair of the frequently associated systemic and pulmonary venous connection anomalies and right ventricular outflow tract obstruction. Because of this high degree of complexity, functional single ventricle palliation has been recommended as a treatment option for DORV-AVSD by some groups. However, biventricular repair in these patients, despite its complexity, can be accomplished with excellent results. Reoperation is common in this difficult group of patients, most commonly for conduit replacement. Reintervention for recurrent left ventricular outflow tract obstruction or atrioventricular valve stenosis or regurgitation is uncommon. We feel that biventricular repair of DORV-AVSD is the preferred treatment option, although further studies are needed to define long-term outcomes.

Keywords Double outlet right ventricle • Complete atrioventricular septal defect • Complete atrioventricular canal • Endocardial cushion defect • Biventricular repair

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Introduction

Atrioventricular septal defect (AVSD) and double outlet right ventricle (DORV) each have a 0.2 % prevalence amongst all live births [1]. Surgical management of these defects has improved over the past few decades such that low mortality and morbidity is expected of repair of major variations of these defects [2–6]. The combination of double outlet right ventricle and atrioventricular septal defect (DORV-AVSD) is rare. In a pathological series of over 500 AVSD specimens, 6.7 % also had DORV [7]. Despite its rare occurrence, DORV-AVSD is a challenging defect to repair and has seen an evolution to its management over the past few decades. It has been more than four decades since it had been postulated that DORV-AVSD could be repaired surgically by enlarging the ventricular septal defect and combining standard repairs for the other lesions [8]. Successful repair of DORV-AVSD was first reported in 1980 [9], but the mortality was high, especially in the presence of associated defects, such as pulmonary stenosis, heterotaxy or anomalous pulmonary venous connections [9]. Early reports, as well as some contemporary surgical series of DORV-AVSD, have combined this defect with the lesion TOF-AVSD [9–11]. The inclusion of TOF-AVSD with DORV-AVSD in early series is likely due to the rare incidence, as well as the similar technique that was used to approach both of these lesions surgically, as discussed later in this chapter. Because of this grouping, an in depth compilation of all series of DORV-AVSD is difficult. Other surgical series [6] define DORV-AVSD as only those patients where the great vessels are 200 % supported by the right ventricle (see Chap. 25). We recently described our experience with DORV-AVSD, which represents one of the largest series published to date [12]. We will refer to this cohort of patients extensively throughout the chapter as we discuss its anatomy and treatment. This chapter will review the anatomy of DORV-AVSD, historical aspects of treatment, our current surgical approach and contemporary results.

Anatomy

Double outlet right ventricle with AVSD is a combination of two lesions that, when considered in isolation, have well defined morphological features [13–16] (Fig. 24.1). Double outlet right ventricle is defined by the origin of the majority of both great vessels from the morphological right ventricle and the presence of a muscular subaortic infundibulum or conus. Importantly, this definition of DORV excludes the anatomy typical for tetralogy of Fallot, where aortic override of the right ventricle is less than or equal to 50 % and there is no muscular subaortic infundibulum. In DORV-AVSD, the main component of the VSD is of the inlet type. While there may be some outlet extension of the VSD, there is subaortic infundibular muscle which separates the defect from the aortic valve annulus. Therefore the VSD in DORV-AVSD can be considered to be remote and non-committed [12]. The

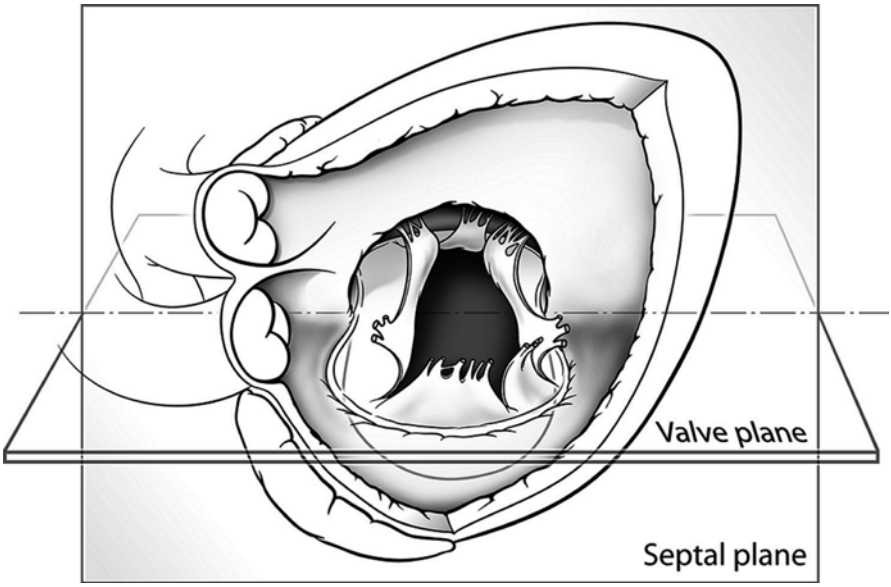

Fig. 24.1 Anatomy of DORV-AVSD with orthogonal atrioventricular valve and septal planes (Used with permission from Devaney et al. [12], with permission of Elsevier)

spatial relationship of the arterial trunks with one another can take any form as in isolated DORV: side by side as well as aorta anterior and to the right or left of the pulmonary artery (PA) [17]. In our series of DORV-AVSD patients, the great arteries were arranged in side-by-side fashion in 4 (with the aorta to the right), positioned with the aorta anteriorly and to the right in 6, and with the aorta anteriorly and to the left in 6 [12].

The definition of an AVSD is a common atrioventricular junction guarded by a common valve. These can be classified according to the Rastelli classification as A, B or C, which is based on the anatomy of the superior bridging leaflet. In our series, as well of those of others, nearly all patients had Rastelli type C [6, 12, 18]. Important atrioventricular valve regurgitation may also be present, and pulmonary stenosis occurred in the majority of our patients with DORV-AVSD [12].

Since DORV-AVSD is associated with heterotaxy syndrome, other cardiac lesions are common. Approximately two-thirds of our patients with heterotaxy had right atrial isomerism [6]. This diagnosis was made either preoperatively or intraoperatively using a number of criteria based on splenic anatomy, bronchopulmonary anatomy, venoatrial connections and morphology of the atrial appendages [19, 20]. Cardiac features of heterotaxy syndrome that may be found in association with DORV-AVSD include anomalous hepatic venous return, total anomalous pulmonary venous connection (TAPVC), interrupted inferior vena cava (IVC), bilateral superior vena cavae (SVC), partial anomalous pulmonary venous connection, common atrium, right aortic arch, and intestinal malrotation. These associated anomalous systemic and pulmonary venous connections further complicate the repair of

DORV-AVSD. Furthermore, none of the patients in our series of DORV-AVSD had trisomy 21.

Diagnosis and Imaging

The diagnosis of DORV-AVSD is made with transthoracic echocardiography. Essentially all anatomical features must be delineated prior to considering repair. These include venous connections, atrioventricular valve structure and function, right and left ventricular size and function, VSD anatomy, and great artery spatial relationship and position within the right ventricle. Cardiac magnetic resonance imaging can provide further anatomical details not adequately defined by echocardiography.

Checklist for Surgical Repair

The inherent complexity and the myriad of associated lesions that must be repaired during repair of DORV-AVSD makes a detailed, specific checklist for the surgeon important, as many physiologic and anatomic details need to be considered. Many of these specific details can be determined by transthoracic echocardiography and cardiac magnetic resonance imaging. Listed below are necessary issues that must be considered prior to embarking on a biventricular repair of DORV-AVSD.

1. Prior palliative operations: BT shunt or pulmonary artery band
2. Atrial inflow anomalies: systemic and hepatic venous anatomy
3. Ventricular size
4. Atrioventricular valve anatomy: mural leaflet size, papillary muscle anatomy.
5. Z scores for atrioventricular and semilunar valves
6. Associated anomalies: TAPVC, intestinal malrotation.

Surgical Approach

Our method of DORV-AVSD repair can be viewed as a 2-patch method of AVSD repair that has been modified to account for abnormalities in systemic and pulmonary venous connection as well as an absence of a contiguous left ventricular outflow tract (LVOT) that connects the left ventricle to the aorta. The septation of the common atrioventricular valve follows the two-patch method that we employ for isolated complete AVSD repair to allow for the creation of two competent, nonstenotic right and left atrioventricular valves [2]. However, modifications are made to the reconstruction of the atrial septum and shape as needed for anomalies of venous

connection, as well as the contour and position of the VSD patch to account for the location of the aorta as it arises from the right ventricle. Further, pulmonary outflow obstruction may need to be addressed.

Biventricular repair of DORV-AVSD requires cardiopulmonary bypass, which is instituted via bicaval and ascending aortic cannulation with moderate hypothermia. Deep hypothermia with low-flow cardiopulmonary bypass or transient hypothermic circulatory arrest may be utilized during the repair of pulmonary venous connection anomalies to improve visualization. After cardioplegic arrest, a right-sided atriotomy is performed to provide access to the atria and common atrioventricular valve. In some anatomic varieties associated with heterotaxy, the approach to the internal cardiac structures may be more appropriate through a left-sided atriotomy.

Repair of the AVSD is then performed. The initial steps of the repair are identical to the ones used in the two-patch method for isolated complete AVSD [2]. The intracardiac anatomy is inspected, and special attention is first given to the atrioventricular valve coaptation points. The inlet component of the ventricular septal defect is then partially closed. A crescent shaped patch of polytetrafluoroethylene (PTFE) is fashioned and sewn to the crest of the defect. This is accomplished by first starting the suture line at the midpoint of the ventricular septum and corresponding posterior portion of the patch and continuing it inferiorly and posteriorly to the junction of the muscular ventricular septum and the annulus of the atrioventricular valve (Fig. 24.2). The entire anterior and superior half of the patch is left unsutured. Marking sutures are then placed in [1] the patch superiorly where it would join the atrioventricular valve annulus and [2] at the midpoint of the inlet component of the ventricular septal defect (Fig. 24.2).

At this point the repair proceeds in an identical manner to a complete AVSD repair. The corresponding portions of the superior and inferior bridging leaflets are secured to the crest of the patch. This is accomplished by placing a series of mattress sutures through the crest of the patch and then passing them through the hinge points of the atrioventricular valve. These sutures are left untied, as they will also be used to anchor the patch used to repair the atrial component of the defect. The zone of apposition between the left ventricular components of the superior and inferior bridging leaflets or "cleft" is then closed with fine interrupted sutures. Next the atrial septum is reconstructed using an autologous pericardial patch. In heterotaxy syndrome, associated bilateral SVC, separate hepatic vein and IVC connections, and anomalies of the pulmonary venous connection can make the reconstruction of the atrial septum challenging with a single patch. Thus, in these situations, separate pericardial patches may be required to fashion the complex baffle geometry without obstruction.

At this stage of the repair, the superior portion of the VSD remains unsutured and will be used to fashion the pathway between the left ventricle and the aortic valve. The next stage of the procedure is focused on the creation of an unobstructed left ventricular outflow tract (LVOT) and establishing RV to PA continuity, if needed. Accessing the subaortic region where the new LVOT will be constructed is best approached through a ventriculotomy (Fig. 24.3) made in the free wall of the right ventricle. The previously placed marking sutures are transposed from the atrium through the ventriculotomy.

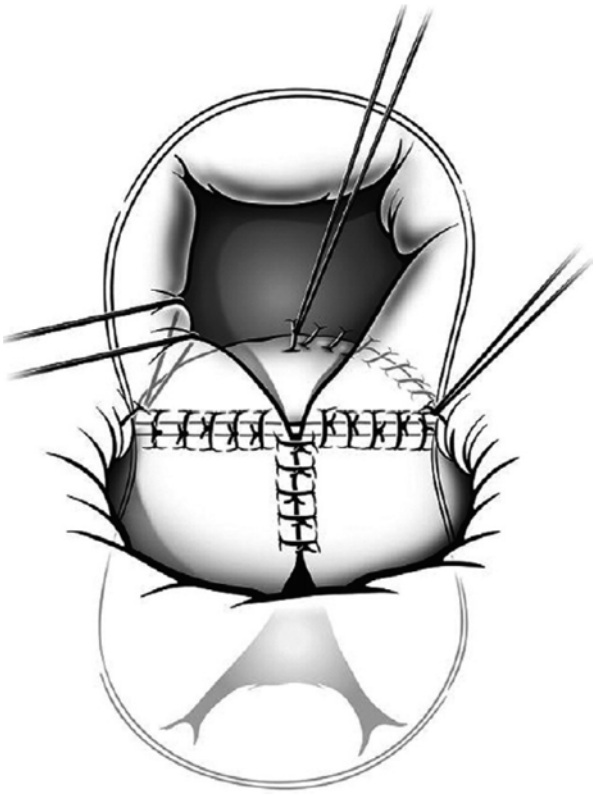

Fig. 24.2 Biventricular repair of DORV-AVSD starts with repair of complete AVSD component, leaving the superior portion of VSD patch unsutured (Used with permission from Devaney et al. [12], with permission of Elsevier)

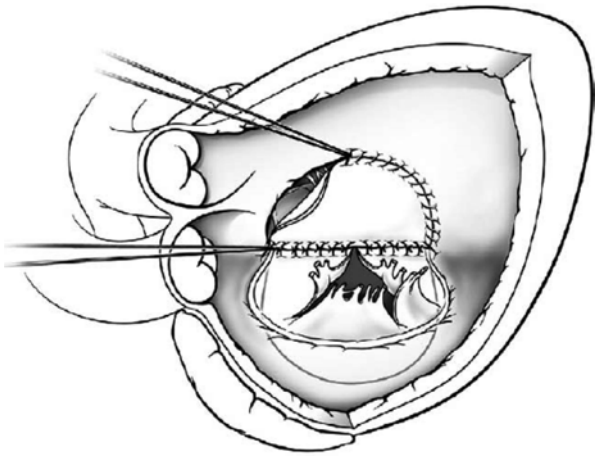

Fig. 24.3 After the atrial septum is reconstructed, a right ventriculotomy is made to provide exposure to the muscular outlet septum and superior portion of VSD patch (Used with permission from Devaney et al. [12], with permission of Elsevier)

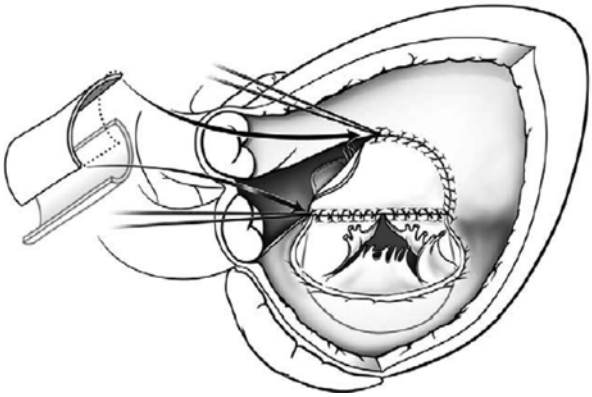

Fig. 24.4 The muscular outlet septum is resected from the superior portion of VSD patch to the aortic annulus and the left ventricular outflow tract is reconstructed with a PTFE patch (Used with permission from Devaney et al. [12], with permission of Elsevier)

The ventricular septal defect usually needs to be enlarged because it is generally confined primarily or entirely to the inlet septum, making it remote from the aortic valve. In our recent series, the VSD was enlarged in 11/16 patients [12]. Attempting to baffle such remote defects to the aortic valve without enlarging the ventricular septal defect is likely to result in obstruction of the left and right ventricular outflow tract as well as interference with the right atrioventricular valve apparatus. Enlarging the VSD in the direction of the subaortic region essentially shortens the LVOT, thus addressing the above pitfalls.

Enlarging the VSD is accomplished by resecting the muscular outlet or infundibular septum. This resection spans the region between the unsutured edge of the prosthetic patch/superior portion of the crest of the ventricular septum and the subvalvar area of the aortic valve (Fig. 24.4). Closure of the remaining portion of the ventricular septum to create an unobstructed LVOT is then undertaken. A second patch of PTFE is trimmed from a tube graft (Fig. 24.4), and then sutured to the free edge of the original patch. The suture lines are then carried superiorly along the edges of resected septum up to the aortic valve annulus (Fig. 24.5).

As a final step, the right ventricular outflow tract is reconstructed. In patients with pulmonary stenosis or atresia, division of septoparietal trabeculations in the right ventricular outflow tract is performed, or a conduit may be placed from the right ventricle to the pulmonary arteries. In our series of 16 patients, 13 received a conduit and one patient received a right ventricular outflow patch with a monocusp valve. Conduits used include bovine jugular venous xenografts, pulmonary allograft, aortic allograft and porcine heterograft.

After separation from cardiopulmonary bypass, the repair is assessed by direct pressure measurements to determine left and right ventricular outflow and, if necessary, atrial inflow gradients. Intraoperative transesophageal echocardiography is performed to assess the atrial and ventricular inflow and outflow pathways, atrioventricular valve stenosis and regurgitation and semilunar valve function.

Other reported series [6] of repair for DORV-AVSD have utilized an approach shared with the repair of TOF-AVSD. This method utilizes a single VSD patch that

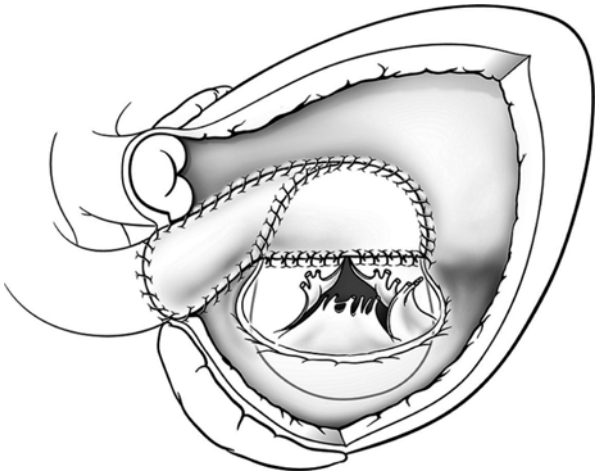

Fig. 24.5 Completed reconstruction of LVOT and complete AVSD with two separate PTFE patches. The right ventricular outflow tract is reconstructed with an outflow patch or valved conduit (Used with permission from Devaney et al. [12], with permission of Elsevier)

is comma-shaped with the wider end of the patch forming part of the LVOT [9, 21]. It has been recommended that the superior portion of the patch should be larger in cases where the aorta is located more rightward of the ventricular septum [21]. We view the repair of DORV-AVSD as being distinctly different from that for TOF-AVSD primarily because of the exclusive right ventricular origin of the aorta, with the aortic valve supported by a distinct muscular infundibulum. This anatomical arrangement makes redirecting the outflow from the left ventricle through the VSD to the aortic valve difficult and prone to obstruction. We and others feel that even when the communication opens toward the outlet region, it is frequently difficult to avoid obstruction of the newly created channel from the left ventricle without enlarging the VSD, also referred to as "VSD translocation" [22].

There have been a few early reports of a physiological repair of DORV-AVSD utilizing a double-switch procedure in which the morphologic right ventricle remains as the systemic ventricle and the morphological left ventricle remains connected to the pulmonary arteries via a conduit, although in the current era it is recognized that this arrangement is not preferable [9, 23]. Because of the risk LVOT obstruction and the complexity of biventricular repair, some groups have recommended a functional single ventricle palliation [24–27]. However, cautious enthusiasm for this strategy must be exercised because many patients may be poor candidates owing to pulmonary artery stenosis, pulmonary hypertension, pulmonary venous obstruction, atrioventricular valve insufficiency, as well as the well-recognized long-term complications of the Fontan procedure.

Results

We have recently summarized our contemporary experience at the University of Michigan C. S. Mott Children's Hospital Congenital Heart Center. In our series, 16 patients were described as having a biventricular repair of DORV-AVSD. The

median age at the time of operation was 16 months. A complete repair was accomplished as the primary operation in only 6 patients. The remainder underwent at least one palliative procedure and/or repair of an associated lesion. Seven patients with inadequate pulmonary blood flow required a systemic-to pulmonary shunt. Three of these patients also underwent concomitant repair of total anomalous pulmonary venous connections. One patient underwent banding of the pulmonary trunk to restrict pulmonary blood flow. One patient who was previously shunted underwent a patch augmentation of the branch pulmonary arteries. Isolated repair of total anomalous pulmonary venous connection was performed in 2 patients. One patient who had undergone repair of infracardiac total anomalous pulmonary venous connection developed pulmonary venous stenosis which was addressed at the time of biventricular repair. In one patient, single ventricle palliation had been previously performed by means of a total cavopulmonary connection at another institution.

Biventricular repair was successfully accomplished in all 16 patients. The median cardiopulmonary bypass time was 242 min, with a median cross-clamp time of 158 min. Arrhythmias were common. Most of these were transient tachyarrhythmias with 8 patients having junctional ectopic tachycardia. Complete heart block requiring a permanent pacemaker developed postoperatively in 2 patients. One patient required ECMO postoperatively because of severe hypoxemia. Another patient had a prolonged recovery due to ventilator dependence and hydrocephalus. There were two deaths in our series. There was one early death that was secondary to pulmonary vascular obstructive disease and one late death secondary to complications related to renal failure.

Five patients had moderate atrioventricular valve insufficiency at discharge while no patient had stenosis. None had significant residual ventricular septal defects or left ventricular outflow tract obstruction. At a median follow-up of 66 months, all discharged patients were clinically well and in New York Heart Association class I or II heart failure.

Causes of reoperation in isolated DORV or CAVSD repair are shared in the biventricular repair DORV-CAVSD, and reoperation is not uncommon in these patients. In our series, six patients needed reoperation. A patient who initially received a pulmonary monocusp valve required a pulmonary valve replacement and atrioventricular valve repair 8 months after repair. Another patient required replacement of a left atrioventricular valve and pacemaker implantation 3 months after repair. Right ventricle to pulmonary artery conduit replacement was required in two patients. Another patient had a significant residual atrial septal defect from a patch dehiscence which required reoperation. The rate of reoperation in our series is not different from those that have been reported by other groups [11].

Three patients have at least moderate atrioventricular valve regurgitation. None have experienced obstruction of the left ventricular outflow tract. Total anomalous pulmonary venous connection was the only significant risk factor for mortality and significant morbidity in our series.

Because DORV-AVSD is such a rare lesion, the optimal surgical approach will be difficult to determine. Nevertheless, we feel that our results support biventricular repair, even in the presence of complex venous connections and a remote interventricular communication. Biventricular repair is associated with a significant rate of reoperation, particularly for replacement of the conduit, but this may ultimately provide long-term

benefits when compared with single ventricle palliation. Formal clinical trials and collaborative investigative groups that provide a comprehensive comparison of these two approaches for DORV-AVSD are needed to evaluate long-term outcomes.

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