Chapter 23
Bi-ventricular Repair of Double Outlet Right Ventricle

Francois Lacour-Gayet

Abstract Double outlet right ventricle (DORV) is a type of ventriculo-arterial connection where the great vessels arise entirely or predominantly from the right ventricle. The name DORV encloses a vast spectrum of congenital heart diseases with various clinical presentations and surgical treatment. Rarely a congenital heart disease has been the source of such controversial exchanges around the anatomical definitions and the optimal surgical techniques. The recent Functional Classification has greatly simplified the management of DORV in describing different functional types unified by a common clinical presentation and more importantly by a similar surgical management. The surgical repair of DORV-VSD type, DORV-Fallot and DORV-TGA (Taussig-Bing) is today performed with very low mortality. The surgical anatomy and the optimal surgical technique of DORV non-committed VSD and DORV-AVSD-Heterotaxy are still controversial. In presence of two viable ventricles, we believe that biventricular repair is the optimal solution. This chapter and Chaps. 15 and 24 describe in details biventricular repair of DORV.

Keywords Congenital heart disease • Double outlet right ventricle • Conotruncal anomaly • Cardiac surgery • Anatomical repair • Pathology • Echocardiography • Pediatrics

Introduction

DORV remains the source of many controversies in anatomical definition and surgical management. DORV, a typical conotruncal anomaly, is not a single disease but essentially a primitive form of ventriculo-arterial connection with great variations in anatomy, clinical presentation and surgical management. DORV is classically classified according to the relationship of the VSD with the great vessels, as defined by

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Lev et al. [1]. The surgeons, in charge of patients with DORV, have adopted for their great majority a Functional Classification of DORV [2], which has clarified the surgical indications of this complex condition.

Surgical Anatomy of DORV

The surgical anatomy is focusing on specific morphological structures relevant to surgical technique.

Anatomical Definitions

DORV is a primitive (Chaps. 2, 3, and 39) form of ventriculo-arterial connection where the two great vessels arise entirely or predominantly from the right ventricle. This conotruncal anomaly presents with various cardiac "phenotypes" and frequent extra cardiac anomalies. DORV is characterized by a large spectrum of lesions depending on: – the location of the VSD, – the relationship of the VSD and the great vessels, – the presence of pulmonary outflow obstruction and – the size of the ventricles. The anatomical definition of DORV remains controversial. There is not one but several definitions of DORV currently used.

- Historically, it is A. Calhoun Witham [3] who coined in 1957 the term double outlet right ventricle and defined it as a "partial transposition complex" by opposition to complete TGA. This definition was later endorsed by Neufeld et al. [4, 5].
- Lev et al. [1] produced in 1972 the universally accepted anatomical definition of DORV, based on the relationship of the VSD with the great vessels.
- Richard Van Praagh defines DORV as a fibrous discontinuity between the mitral and aortic valves due to the persistence of a subaortic conus [6, 7]
- Robert H. Anderson established the popular "50 % rule" [8, 9], which states that a DORV is present when more than 50 % of both arterial valves are connected to the morphologically right ventricle.
- R. Sakata and Y. Lecompte [10] defines DORV as a malposition of the great vessels
- François Lacour-Gayet et al. [11–13] supports the "200 % rule". From a surgical standpoint, "complete DORV" have both arterial valves arising entirely from the morphologic right ventricle. Consequently, DORV can be called a "Transposition of the Aorta", a synonym of "Partial Transposition"; the first definition given by Witham [3]

Morphology of DORV

DORV is essentially a conal malformation. In the most typical forms, due to the persistence of a subaortic conus [6, 7], DORV is characterized by a double conus, formed by a normal subpulmonary conus and an abnormally present subaortic

conus. The two conus are formed by a common inter-conal septum (conal/infundibular/outlet septum) and their lateral walls. The right lateral conal wall (subpulmonary conus for R. Van Praagh) is normally developed creating a muscular discontinuity between the tricuspid and the pulmonary valve (ventricular infundibular fold, VIF, for RH Anderson). The persistence of the left lateral conal wall (left VIF for RH Anderson) maintains the aorta entirely or partially inside the cephalhead part of the right ventricle, creating an additional subaortic infundibulum with muscular mitro-aortic discontinuity. The conal septum remains inside the right ventricular cavity instead of its normal position between the limbs of the trabecular septo marginalis (TSM). The position of the parietal band (crista supra ventricularis) is variable in DORV. In many instances it can be normally situated in contact with the septal band (TSM). Elsewhere, it can be fairly recognizable or even absent.

The conal septum could be malaligned anteriorly or posteriorly creating a subpulmonary stenosis or a subaortic obstruction. It can be absent or deficient. The left subaortic conus could be more or less developed, with a large or shallow mitro-aortic discontinuity.

The aorta is either: – in normal position, located posteriorly and to the right of the pulmonary artery or – side by side to the right or – located anteriorly and to the right of the PA in the most typical DORV. *The more anterior the aorta, the more complex form of DORV.* When associated with an atrio-ventricular discordance, the aorta is located to the left of the PA.

The VSD in DORV can be located above and in between the limbs of the TSM or below the posterior limb of the TSM. Rarely, it can be situated in the inlet or trabecular muscular septum. It can be absent. The VSD in "complete DORV" is the only outlet of the left ventricle. Any restriction of the VSD prior or following surgery will induce an LV obstruction. The name to give to this "hole" is controversial: – bulbo-ventricular foramen, AV canal type VSD, inlet muscular VSD for Van Praagh [6, 7] or – perimembranous VSD, interventricular communication for Anderson [8, 9] or – primary interventricular foramen for Van Mierop [14–16]. The main point is the location of the conduction tissue in the DORV non committed VSD as discussed further.

It is noticeable that the conal morphology is quite stable in the "complete" form of DORV with two infundibulum, the conal septum standing in the RV, distant from the Y of the TSM and 200 % of great vessels in the right ventricle. In "partial" forms, when the vessels do not arise entirely from the right ventricle, the subaortic conus and its left lateral can be deficient.

Lev-Bharati Classification

Four anatomic types are recognized in Lev and Bharati *anatomical classification* [1] based on the relationship of the VSD with the great vessels:

- DORV with subaortic VSD
- DORV with doubly committed VSD
- DORV with subpulmonary VSD (Taussig-Bing anomaly)
- DORV with uncommitted or remote VSD

Associated Lesions

Associated anomalies are frequent:

- *Ventricular hypoplasia* is the most frequent association. DORV is often a component of single functioning ventricle; a clear contra-indication to biventricular repair
- *Valvar or subvalvar pulmonary obstruction* is frequent; however it is rare in Taussig Bing. Pulmonary atresia is possible.
- A *restrictive VSD* is frequent and should be recognized before surgery. Following biventricular repair, a restricted VSD (diameter less than the aortic valve) will create a subaortic obstruction. The VSD was found restrictive in 27 % of all DORV and in 66 % of DORV-non-committed VSD [11–13, 17].
- *Coarctation and interrupted aortic arch* are associated with the Taussig Bing anomaly [18].
- *Subaortic obstruction* is due to a stenosis on the persitant subaortic conus/infundibulum and is seen in Taussig-Bing and in DORV-nc-VSD.
- A *complete atrioventricular septal defect with pulmonary stenosis and heterotaxy/isomerism* is a complex association. It could be associated with total abnormal pulmonary venous return (TAPVR) and intestinal malrotation [13]
- *Pulmonary atresia and pulmonary arteries branches stenoses* can be present.
- *Straddling AV valves* is a complex association and could be a contra-indication to bi-ventricular repair
- *Pulmonary hypertension* should be prevented and is evaluated in patients seen late
- *AV discordance* can be associated with DORV [9].
- Other *rare associations* include: situs inversus, ectopia cordis, etc…
- *Chromosomic anomalies* are present in 20 % of DORV.

Functional Classification

Due to the variety of anatomic forms and the absence of consensus on the definition, the Functional Classification was created to clarify the surgical indications. It was adopted together by the Society of Thoracic Surgeons (STS) [2], the European Association of Cardiothoracic Surgery (EACTS) and by the Association for European Pediatric Cardiology (AEPC). It is based on the relationship of the VSD with the great vessels and the presence of a pulmonary outflow obstruction. These factors allow describing different *functional types* unified by a similar clinical presentation and more importantly by a common surgical management.

The Functional Classification of DORV recognizes four types [2]. The author is adding DORV-CAVSD as a fifth type (Fig. 23.1).

1. DORV-VSD type (DORV subaortic VSD and DORV doubly committed VSD)
2. DORV-Fallot type
3. DORV-TGA type. Taussig-Bing
4. DORV remote VSD
5. DORV-CAVSD-PS-Heterotaxy

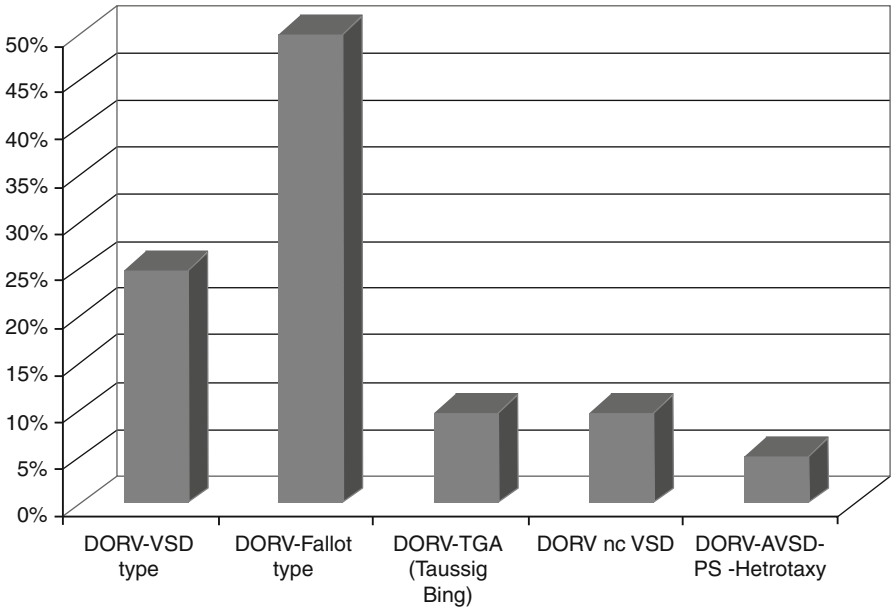

Fig. 23.1 The Functional Classification of DORV recognizes four types. The author is adding DORV-CAVSD as a fifth type. DORV non committed VSD and DORV-AVSD are complex DORV and require original and challenging surgical techniques, specific to this condition

DORV non committed VSD and DORV-AVSD are complex DORV and require original and challenging surgical techniques, specific to this condition.

DORV-VSD Types. They account for 25 % of DORV (Fig. 23.1) and Include Two Different Forms: DORV Subaortic VSD (Fig. 23.2) and DORV Doubly Committed VSD (Fig. 23.3)

DORV- Subaortic VSD (Fig. 23.2)

The VSD is located above and in between the limbs of the trabecula septo marginalis (TSM). There is no pulmonary stenosis. The clinical presentation is that of a VSD with large left-to-right shunt. The VSD can be restrictive [13, 17]. The aorta rarely arises entirely from the right ventricle. We considered this form as a "partial" DORV (not always 200 %).

DORV Doubly Committed VSD (Fig. 23.3)

This form seems the most infrequent one, around 3 %. When the VSD is doubly committed, the conal septum is absent or markedly deficient. Sometimes, the VSD

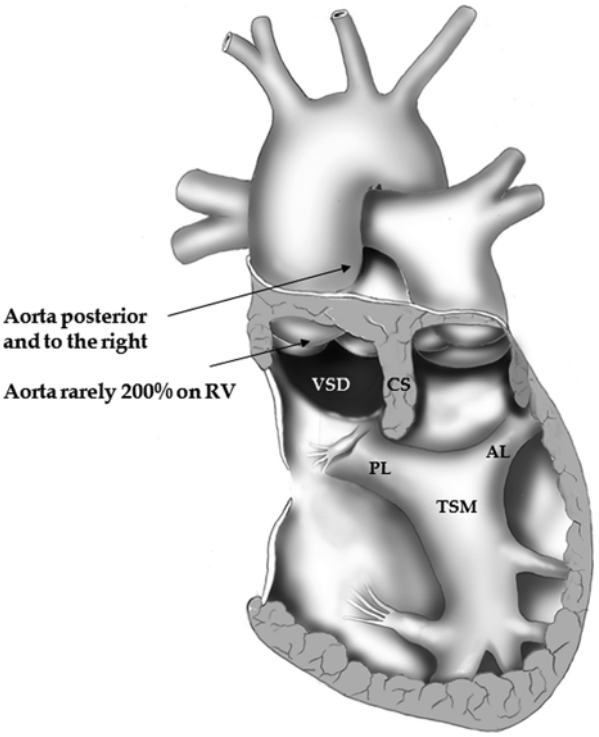

Fig. 23.2 DORV subaortic VSD. The VSD is subaortic, standing above the TSM. The aorta is in normal position posterior and to the right. It is not always arising entirely (200 %) from the right ventricle. The subaortic conus is rarely well developed (*VSD* ventricular septal defect, *TSM* trabecula septo marginalis/septal band, *AL* anterior limb, *PL* posterior limb)

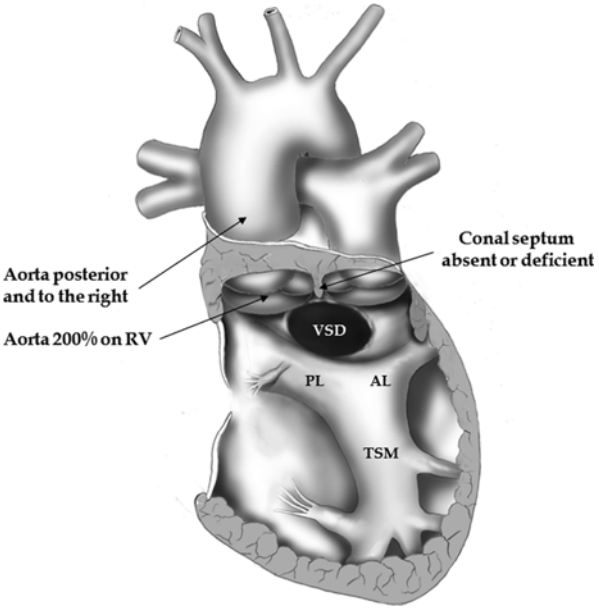

Fig. 23.3 DORV doubly committed VSD. The conal septum is absent or deficient. The VSD is located below the two arterial valves and above the TSM. The aorta is usually in normal position. Many times, the aorta is arising entirely (200 %) from the right ventricle (*VSD* ventricular septal defect, *TSM* trabecula septo marginalis/septal band, *AL* anterior limb, *PL* posterior limb, *CS* conal septum)

is located medially underneath the two orifices. Elsewhere, the VSD is located below the aorta or below the pulmonary, being a transitional form of DORV subaortic VSD or DORV-Taussig Bing. The aorta is most often entirely located inside the right ventricle, sitting side by side and to the right of the PA. A cyanosis could be present.

DORV-Fallot Type (*Fig. 23.4*)

In DORV-Fallot type, the VSD is subaortic and a pulmonary stenosis is constantly associated. With an occurrence around 50 %, it is the most common type. Patients with DORV-Fallot type present like a Tetralogy of Fallot. The degree of cyanosis depends on the severity of the pulmonary stenosis. The VSD can be restrictive in 20 % [17]. The aorta rarely arises entirely from the right ventricle. It is difficult to differentiate DORV-Fallot type from tetralogy of Fallot, and the diagnosis of DORV is frequently overestimated.

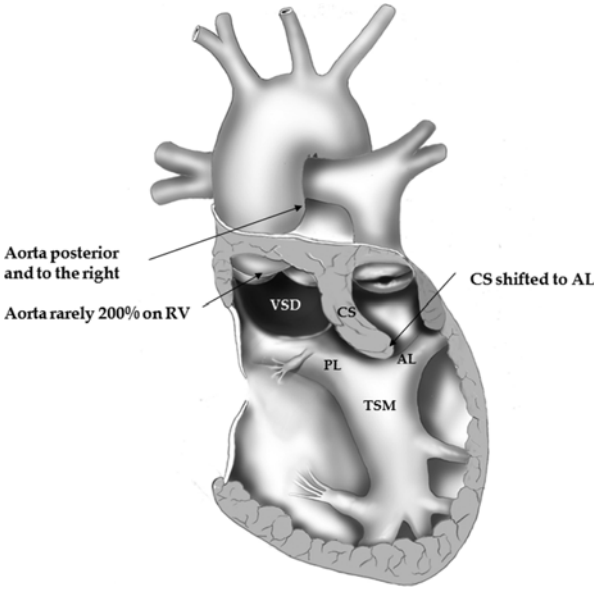

Fig. 23.4 DORV-Fallot. The VSD is subaortic, located above the TSM. The conal septum is shifted toward the anterior limb of the TSM, creating a pulmonary flow obstruction. The aorta is in normal position, posterior and to the right. It is not always arising entirely (<200 %) from the right ventricle. The subaortic conus is partially developed (*VSD* ventricular septal defect, *TSM* trabecula septo marginalis/septal band, *AL* anterior limb, *PL* posterior limb, *CS* conal septum)

A rare variant is DORV-Fallot with doubly committed VSD, where the conal septum is reduced or absent and the pulmonary annulus restrictive. It is similar to the Asian form of TOF.

DORV-TGA Type. Taussig-Bing (Fig. 23.5)

Taussig-Bing represents 10 % of DORV (see Chaps. 14 and 15). The clinical presentation is similar to patients with TGA-VSD. Aortic arch obstruction and subaortic obstruction are frequently associated. The great vessels are side by side, with the aorta on the right. The coronary arteries have usually a double looping pattern (Chaps. 14 and 15). The RV can be small but not really hypoplastic in neonates. The VSD is exceptionally restrictive. The aorta arises entirely from the right ventricle and the pulmonary artery arises more than 50 % from the left ventricle but rarely entirely. There are two conus and the conal septum is shifted toward the posterior limb of the TSM, creating potentially a subaortic obstruction (Fig. 23.5). It is often difficult to differentiate Taussig-Bing from TGA-VSD, depending on the limits chosen to anatomically

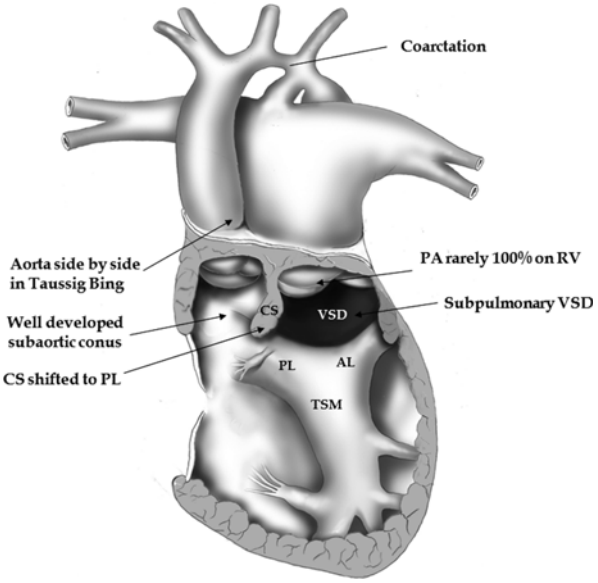

Fig. 23.5 DORV-TGA, Taussig Bing. The VSD is subpulmonary (Taussig Bing), located above the TSM. The conal septum is shifted toward the posterior limb of the TSM, creating potentially a subaortic obstruction. There is frequently a coarctation. The aorta is side by side and to the right of the pulmonary artery. The subaortic conus is well developed. The aorta is arising entirely from the right ventricle, while the PA arises most often only partially from the right ventricle [19]. In Taussig-Bing, there is a mitro-pulmonary discontinuity (*VSD* ventricular septal defect, *TSM* trabecula septo marginalis/septal band, *AL* anterior limb, *PL* posterior limb, *CS* conal septum)

separate the two ventricles; -either the axis of the muscular septum [6] or the interventricular communication [9]. The Taussig-Bing anomaly should have mitro-pulmonary discontinuity [19, 20], whereas the TGA-VSD have mitro-pulmonary continuity.

DORV-Non-committed VSD *(Figs. 23.6 and 23.7)*

This complex form of DORV, accounts for 10 % and presents with or without PS. Both great vessels arise entirely from the right ventricle (200 %) and this is the most typical form of "complete DORV", as shown on echocardiogram or angiogram (Fig. 23.6). The aorta stands anterior and to the right of the PA. The aorta is very remote from the VSD, by a distance described as "considerable" by Van Praagh et al. [6]. Belli et al. [21] arbitrarily defined DORV nc VSD, as a DORV where the

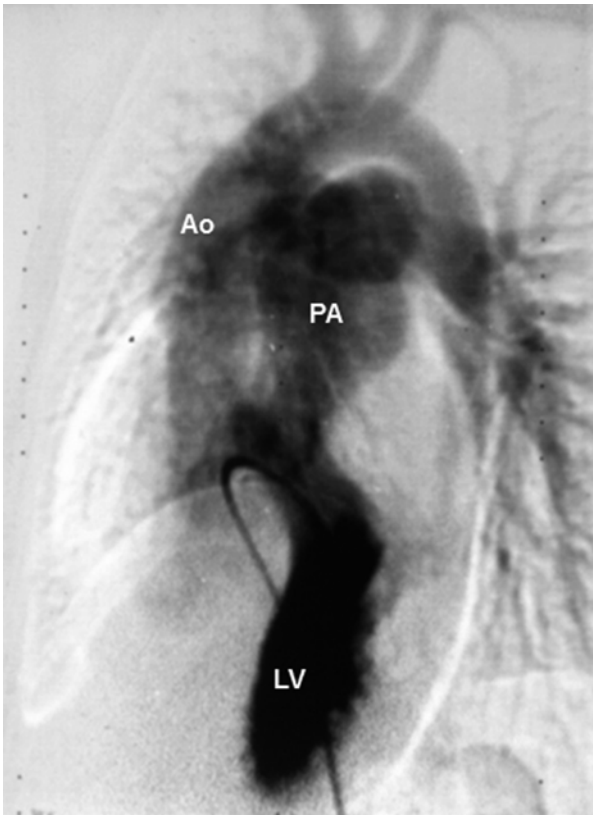

Fig. 23.6 DORV non-committed VSD angiogram. Notice that both great vessels arise 200 % from the right ventricle

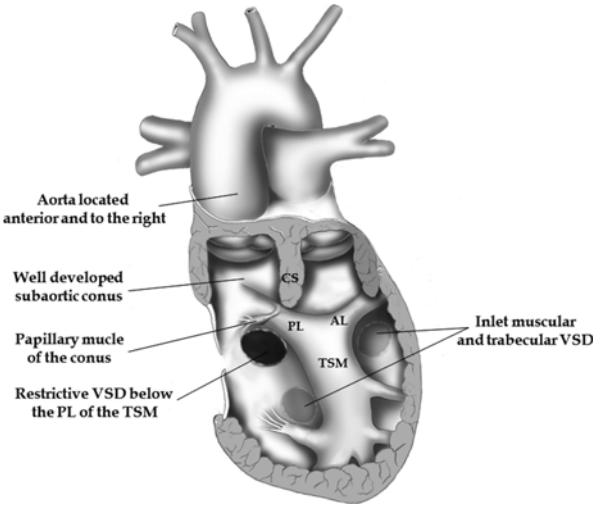

Fig. 23.7 DORV non committed VSD. The VSD is located below the PL of the TSM. It is frequently restrictive. The conal septum could be shifted toward the PL creating a potential subaortic obstruction or toward the AL (not represented on the figure) creating a pulmonary stenosis. The aorta is anterior and to the right. There is nearly constantly a mitro-pulmonary discontinuity. There are clearly two infundibuli with the subaortic conus and conus wall (left VIF for RH Anderson) being very well developed. In rare cases, the VSD can be located in the muscular or trabecular septum. Notice on this drawing that the papillary muscle of the conus is in the way of a tunnelization of the VSD to the aorta (*VSD* ventricular septal defect, *TSM* trabecula septo marginalis/septal band, *AL* anterior limb, *PL* posterior limb, *CS* conal septum, *VIH* ventriculo-infundibular fold)

distance between the superior edge of the VSD and any great vessel annulus is greater than the aortic valve diameter. The VSD is usually in contact with the tricuspid valve (Fig. 23.8) [8, 23]. It is rarely a truly inlet muscular or trabecular VSD (Fig. 23.6) [24]. The VSD is located below the posterior limb of the TSM; different from a Taussig Bing where the VSD is located above the posterior limb of the TSM [24]. In DORV nc VSD, *it is the aorta that is remote not the VSD*. In our experience, *the VSD was restrictive in nearly two third of the patients* [12, 13, 17]. The VSD has a natural tendency to close with time. The VSD can be absent with hypoplasia of the left ventricle [6], or obturated with time following a Fontan operation performed on DORV nc VSD [17, 25, 26]. A closing VSD can also be seen in the neonatal period requiring an urgent decompression of the left ventricle. There is usually an "inverted coronary anatomy" (see Chap. 14). There is *almost constantly a fibro-aortic discontinuity* as well as two well-developed infundibulum. The conal septum stands entirely in the right ventricle. It can be shifted toward the posterior limb of the TSM, potentially creating a subaortic obstruction.

Pulmonary outflow obstruction is present in one third of the cases. This form is different from a DORV-Fallot because the aorta is distant from the VSD by a longer distance, according to our definition [21]. The conal septum is shifted toward the anterior limb of the TSM, which opens up the subaortic infundibulum. Straddling AV valve and other association can be seen.

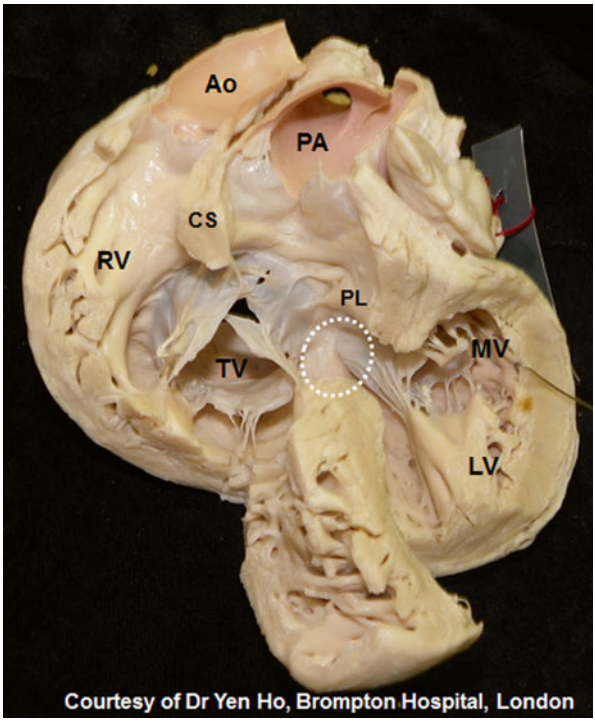

Fig. 23.8 DORV nc VSD Specimen. This specimen from the Morphologic Congenital Heart Disease Department of the Brompton Hospital, London is a "spectacular" DORV nc VSD. The ventricular septum was cut off and the two ventricles are well seen. The VSD is an oval in white. It is located below the TSM and remote from the two arterial valves. It is restrictive. Notice that it is impossible to baffle the VSD to the aorta due to the tricuspid valve and the conal septum. On the other hand, it is possible to tunnelize the VSD to the PA and add an arterial switch (see Fig. 23.11). Notice also that it is the aorta that is remote and not the VSD (*PL* posterior limb, *CS* conal septum, *RV* right ventricle, *LV* left ventricle, *TV* tricuspid valve, *MV* mitral valve) (Courtesy of Dr. Yen Ho, Brompton Hospital, London. Used with permission)

DORV-CAVSD-PS-Heterotaxy *(Figs. 23.9, 23.10, and 23.11)*

DORV-CAVSD is a complex form and accounts for 5 %. Pulmonary outflow obstruction is nearly constant. The clinical presentation is that of a Fallot-AVSD with cyanosis. The VSD has usually a large anterior component like in Rastelli type C (Figs. 23.9, 23.10, and 23.11) [22, 27–29]. The constant presence of heterotaxy makes this condition even more complex due to possible association with TAPVR and intestinal malrotation [13]. Both vessels arise entirely from the right ventricle (200 %) in this complete DORV. This is the best landmark between DORV-Fallot and AVSD-DORV which are difficult to differentiate. Another difference is the absence of Down's syndrome in DORV-AVSD in relation with the presence of isomerism [29]. The bi-ventricular repair of this complex form started 30 years ago [27] with Al Pacifico and is currently achieved by several centers [13, 22, 28–30] (See Chap. 26).

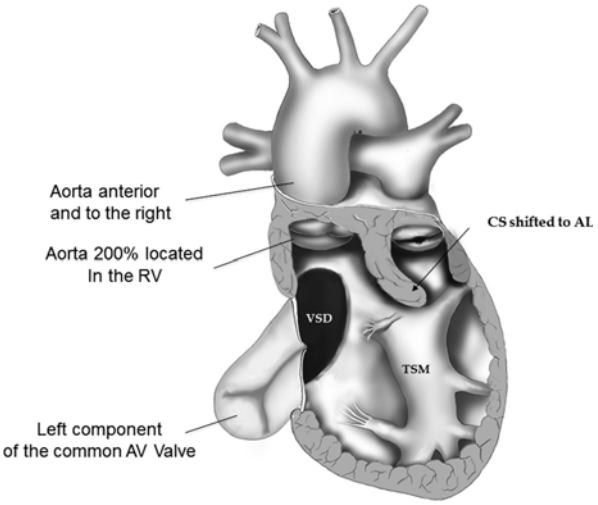

Fig. 23.9 DORV AVSD. The aorta is located anterior and to the right. The great vessels are 200 % on the RV. There is very frequently a pulmonary stenosis with anterior shifting of the conal septum. It is always a type C of Rastelli. The left AV valve is shown. The VSD has an anterior component toward the aorta. The subaortic conus is well developed. The tunnelization to the aorta is usually feasible (see Chap. 23) (*VSD* ventricular septal defect, *TSM* trabecula septo marginalis/septal band, *AL* anterior limb, *PL* posterior limb, *CS* conal septum)

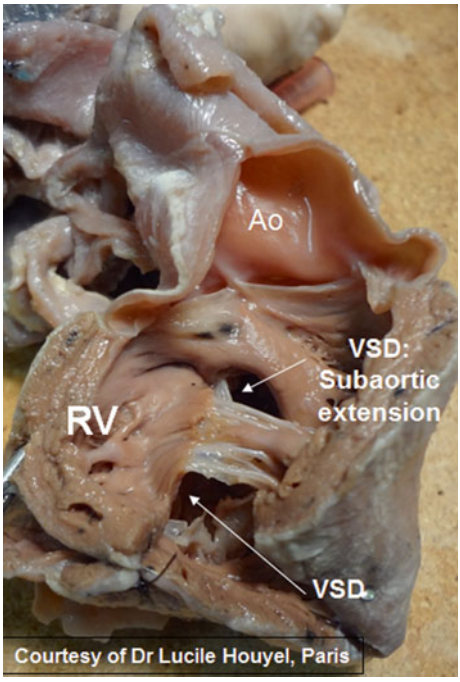

Fig. 23.10 DORV AVSD specimen. This specimen coming from Pathology Department of Marie Lannelongue Hospital, Paris shows clearly the anterior component of the VSD below the aorta (Used with the permission of Dr. Lucile Houyel, Paris) (*VSD* ventricular septal defect, *RV* right ventricle)

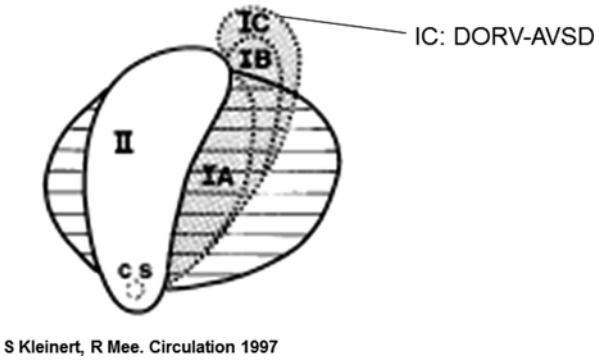

Fig. 23.11 Shape of the VSD in AVSD. *IA* VSD in complete AVSD, *IB* VSD in AVSD-Fallot, *IC* VSD in DORV-AVSD, *II* ostium primum, *CS* coronary sinus (From Kleinert and Mee [22], with permission)

Diagnosis-Imaging of DORV

Echocardiography

An accurate and comprehensive anatomical diagnosis is essential to define the DORV types and the surgical anatomy. *Echocardiography remains the optimal tool* to insure a complete inventory of the lesions. This is a complex evaluation, requiring an experienced operator. It could be useful for the surgeon to attend the exam. The echocardiogram needs to provide several specific information to guide the surgical indications.

The *dimension of the ventricles* will first orient towards a biventricular repair or single ventricle palliation. In borderline ventricular volumes, MRI is indicated.

The *alignment of the great vessels with the ventricular cavities* can be difficult to define. This is visualized with a sweep from the VSD to the great vessels. Depending upon the view-plane used for imaging, the arterial trunks may appear to be connected to different ventricles. It could be extremely difficult to delineate precisely the percentage of arterial valves overriding the right ventricle, even when the two vessels arise entirely from the right ventricle (200 %). The echocardiography could be misleading and the final anatomy ultimately confirmed during surgery. When present, a *mitro-aortic discontinuity* in long axis parasternal view is a major landmark for the diagnosis [6, 7]. In complex forms, the mitro-aortic discontinuity is usually large, whereas in simple forms like DORV-Fallot type or DORV-VSD-type, the mitro-aortic discontinuity could be shallow or absent. A *double infundibulum* is clearly seen in the most typical forms, with a long subaortic infundibulum, which can be narrowed. The other significant landmark in complex DORV is the *conal septum entirely located in the right ventricle*.

The *relationship and the distance between the VSD and the arterial trunks.* This allows deciding the type of the VSD re-routing; either to the aorta or to the pulmonary artery, and also defining DORV nc VSD type.

The diameter of the VSD in comparison with the diameter of the aortic annulus. When smaller than the aortic diameter, the VSD is to be enlarged.

The distance between the tricuspid and pulmonary valves [10, 31]. This space is the site of the baffle to be constructed to connect the VSD to the aorta. It is frequently occupied by the conal septum and the conal papillary muscle (Fig. 23.8).

The *coronary artery anatomy* is usually similar to a normal heart, with the right coronary artery arising from the left antero-lateral sinus and the common left trunk rising for the left postero-lateral sinus. This is the coronary pattern in Taussig Bing, named "inverted" in TGA (see Chaps. 15 and 16). Other coronary anomalies like left anterior descending crossing the infundibulum and single coronary can be seen by echocardiogram.

Cardiac association. Right ventricular outflow tract obstruction is the most frequent association. It can be either valvular and/or infundibular or atretic. *Other associations* are diagnosed: multiple VSD, aortic arch obstruction and subaortic obstruction, straddling AV valves, etc…

Catheterization-Angiography

Catheterization is rarely performed in simple forms. It is useful in complex forms (Fig. 23.6) to evaluate: systemic and pulmonary venous return, coronary anatomy, PA branches size and pulmonary vascular resistances in older patients.

CT Scan. MRI. 3D Echo

Magnetic resonance imaging, computed tomography, and three-dimensional echocardiography provide interesting imaging and *are required in complex forms*. MRI is needed to evaluate border line ventricular volumes.

3D Printing and 3D Computed Reconstruction

3D printing and 3D computed reconstruction based on CT scan [32] are under progress and may become the best tool to define the feasibility of rerouting the VSD to the great vessels in complex DORV.

Prenatal Diagnosis and Chromosomic Anomalies

DORV can be diagnosed by fetal echocardiography with a good degree of accuracy [33] when the two vessels arise from the right ventricle. DORV can be associated with chromosomic abnormalities. Di George-deletion 22q11 syndrome is frequent

in patients with Taussig Bing. CHARGE syndrome and other rare chromosomic anomalies are possible. Trisomy 21 is exceptional in DORV-AVSD-heterotaxy. The occurrence of DORV during the embryologic development is related to a genetic dysfunction of the second heart field which regulates the conotruncal formation [34] (See Chaps. 2 and 36).

DORV Pre-operative Checklist

- Dimension of the ventricles
- Ventriculo-arterial alignment
- VSD-great vessels relationship and distance
- VSD diameter
- Distance tricuspid-pulmonary valves
- Great vessels relationship
- Coronary anatomy
- Associations:

  Pulmonary valvar- subvalvar stenosis, atresia
  Additional VSD
  Subaortic obstruction
  Coarctation, IAA
  ASD, PDA
  CAVSD
  Heterotaxy/Isomerism: LSVC, asplenia, azygos continuation
  TAPVR
  Straddling AV valves
  Intestinal malrotation
  Pulmonary hypertension
  Chromosomic anomaly
  Others: situs inversus, AV discordance, etc…

Surgical Techniques of DORV Biventricular Repair

Repair of DORV-VSD Type

DORV Subaortic VSD

Patients with *DORV-VSD and subaortic VSD* (Fig. 23.2) are treated by one stage repair in the first months of life. A prior PA banding is sometimes indicated and is contra-indicated if the VSD is restrictive. The operation is performed through the tricuspid valve. The VSD is found close to the aortic valve that stands partially and

sometimes entirely in the right ventricle. A right ventriculotomy is rarely requested. The VSD can be restrictive requiring a ventral enlargement [17]. The patch is longer than usual to re-route the VSD to the aorta. It is secured using pledgeted separated stitches and continuous sutures.

DORV Doubly Committed VSD

In DORV doubly committed VSD (Fig. 23.3), a right ventriculotomy is needed. The VSD is infundibular. There conal septum is absent or deficient and the aortic and pulmonary annulus could be in continuity. The VSD baffle to the aorta needs to be sutured between the two annuli and should not be too large to avoid a subpulmonary obstruction. A patch enlargement of the ventriculotomy limits the risk of subpulmonary stenosis. When the tunnelization to the aorta is difficult, the tunnelization to the PA followed by arterial switch should be considered.

Repair of DORV-Fallot Type (Fig. 23.4)

Patients with DORV-Fallot type present like Tetralogy of Fallot. A Blalock-Taussig shunt is considered in the first months of life and is indicated in cases of abnormal coronary, severe PA branches hypoplasia and multiple VSD. The operation is sometimes conducted through the atrium, but a right ventriculotomy offers a better approach to both the aortic valve and the VSD borders. Dividing first the parietal band allows a good visualization of the aortic annulus. The VSD patch should be large enough to reach the anterior part of the aortic annulus that is overriding the right ventricle.

Repair of DORV-TGA Type (Taussig-Bing) (Fig. 23.5)

See Chap. 16

Repair of DORV-Non-committed VSD (Figs. 23.7, 23.8, and 23.12)

Biventricular repair of DORV-nc-VSD remains challenging and is performed only by a few centers [11–13, 21, 22, 24, 28, 30, 31] The VSD is restrictive in 60 % of the DORV-nc-VSD [13, 17] and need to be enlarged as shown below.

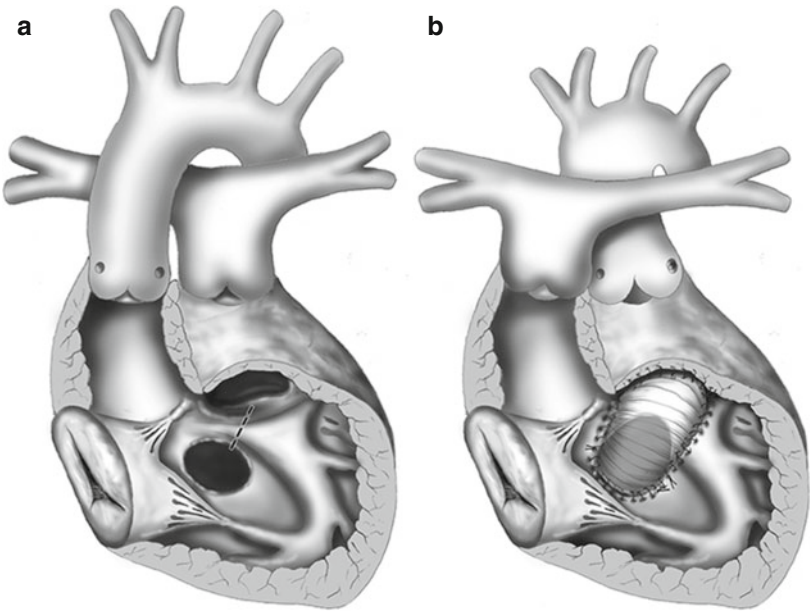

Fig. 23.12 DORV nc VSD, Tunnelization to the pulmonary artery + arterial switch. The VSD is frequently enlarged anteriorly and to the left (**a**). The repair includes a tunnelization to the pulmonary artery and an arterial switch procedure (**b**). Notice that the tunnel is quite short because the tunnel ends on the ostium infundibuli

Repair of DORV-nc-VSD Without Pulmonary Obstruction

A palliative PA banding is preferable due to the need of a significant intraventricular baffle. Depending on the anatomy, the VSD can be tunnelized either to the aorta or to the PA followed by an arterial switch.

Tunellisation of the VSD to the Aorta

This operation is undertaken when the child has reached 6–8 kg. If this technique seems logical, it is in fact quite challenging. In many instances, constructing a tunnel to the aorta requires to resect the conal septum and sacrifice part of the tricuspid valve and/or the papillary muscle of the conus (Fig. 23.8) [10, 24]. The aorta is transposed and always distant from the VSD. It requires a very long patch that could impair the LV function. In these cases, Barbero-Marcial et al. [24] has proposed a multiple patch with resection of the conal septum, reimplantation of the TV chordate on the patch and partial sacrifice of the tricuspid valve or the pulmonary artery.

It is our preference to tunnelize to the VSD to the aorta only when the baffle can be constructed without impairing the conal septum or the tricuspid and pulmonary

valves [11–13]. The baffle is constructed through a double approach, through the tricuspid valve and a right ventriculotomy, using a patch of Goretex or bovine pericardium, sutured with separated pledgeted stitches and continuous suture. When the VSD size is less than the aortic annulus, the VSD is to be enlarged anteriorly as shown on Fig. 23.12.

Tunnelization of the VSD to the PA Followed by an Arterial Switch

This technique was described by the author in 2002 [11] (Fig. 23.12). When the aorta can't be tunnelized to the aorta (Fig. 23.8), it can most often be tunnelized to the PA. This condition is therefore treated like a Taussig-Bing anomaly by tunnelization to the PA, associated with an arterial switch. The only difference being a longer patch and the need to frequently enlarge the VSD. The patch is shorter than the baffling to the aorta because the baffle ends at the level of the ostium infundibuli. In two third of the cases, the VSD needs to be enlarged. The incision of the septum is carried out on the superior border of the VSD and should be large enough as to create a VSD larger than the aortic orifice. Our surgical experience following safe ventral enlargement of the VSD [11–13, 17], suggests that the conduction tissue is located on the inferior rim of the VSD. The conduction tissue could also be located on the superior border into the posterior limb of the TSM and the avoidance of AV block could be due to the early division of the bundle.

The patch is constructed with a triple approach, through the tricuspid valve, the right ventriculotomy and the aorta. The baffle is constructed with pledgeted sutures in the area of septal incision and by continuous sutures. An arterial switch follows. It requires dealing with inverted coronary arteries anatomy, implying to move the reconstructed pulmonary trunk toward the right PA branch (see Chaps. 15 and 16) if a Lecompte maneuver is performed.

Repair of DORV-nc-VSD with Pulmonary Obstruction

In all cases with pulmonary stenosis, *a palliative BT shunt* is preferable.

Tunnelization of the VSD to the Aorta and RVOT Reconstruction

Due to the anterior malalignement of the conal septum toward the pulmonary infundibulum, the space between the tricuspid and the pulmonary valve is often larger, authorizing a safer baffling to the aorta, particularly in pulmonary atresia. This technique of intraventricular repair (not to be confused with R.E.V) is the most adapted to DORV-nc-VSD with RVOTO, providing that the VSD can be safely baffled to the aorta without resection of the conal septum or sacrifice of the PA. The RVOT is simply reconstructed with a transannular patch enlargement or a RV to PA valved conduit in pulmonary atresia.

R.E.V. Operation and Rastelli Procedure

These techniques combined a tunnelization of the VSD to the aorta with an interruption of the pulmonary trunk associated with a RV to PA valved conduit (Rastelli) or a Lecompte maneuver (R.E.V.) [31]. A R.E.V procedure is indicated in patients with restrictive tricuspid- pulmonary valve space, when the resection of the conal septum cannot spare the pulmonary annulus. It includes a resection of the conal septum, the interruption of the PA trunk and a Lecompte maneuver with RV to PA reconstruction using a monocusp valve (see Chap. 20). The conal papillary muscle is frequently detached and reimplanted on the patch. As already mentioned earlier, we are not in favor of impacting on the tricuspid valve apparatus.

Tunnelization to the PA and Arterial Switch

In the rare forms where the pulmonary obstruction is limited to an infundibular stenosis, with a normal PA valve, a tunnelization to the PA with arterial switch could be theoretically performed associated with a patch enlargement of the infundibulum.

Double Root Translocation (see Chap. 21)

This complex operation previously described for TGA-VSD-PS [35, 36] has found indications [37, 38] (see Chap. 21), in DORV nc VSD-PS, where the tunnelization to the aorta is not possible. In DORV-nc-VSD, the two great vessels are entirely located in the RV and the translocation of the aorta on the PA annulus does not repair the DORV. The objective of the double root translocation is only to place the remote aorta, which will remain on the RV, in a position closer to the VSD. This very complex technique is an interesting solution which would need a longer follow up to be adopted in the armamentarium of DORV repair.

Repair of DORV-AVSD-PS-Heterotaxy

See Chap. 24.

Limitations of DORV Bi-ventricular Repair and Fontan Indication

This chapter is dedicated to bi-ventricular of DORV. It is clear that the surgical techniques described above are only indicated in selected patients.

Contra-indication to Biventricular Repair

There are several contra-indications:

- *Hypoplasia of one ventricle* is frequent [6–8]: it is the main contra-indication. Border line left or right ventricular cavities should be assessed by objective measurements of the end-diastolic LV and RV volumes using MRI.
- *Straddling AV valves*. Straddling type C is a contra-indication. It is defined as straddling chordae joining a papillary muscle or the parietal wall of the contro-lateral ventricle. Straddling A and B, when the straddling chordae are located on the edge or at some distance inside the contro-lateral ventricles, can be managed in moving the VSD baffle a few millimeters away [39].
- *Multiple VSD*. More than two associated VSD or an associated apical VSD contra-indicate two ventricles repair [13, 22]
- *Inlet muscular and trabecular VSDs* were described [8, 23, 24] in DORV nc VSD (Fig. 23.7). They are contra-indication to biventricular repair. We have not found such true inlet muscular or trabecular VSD in our series of bi-ventricular repair of DORV nc VSD [11–13].
- *Learning curve*. Bi-ventricular repair of simple forms (DORV-VSD, DORV-Fallot) and of Taussig Bing are routinely performed in most centers. Repair of complex forms is challenging and is considered too risky for many institutions not ready to engage in a learning curve.

Univentricular Repair in Complex DORV

Univentricular repair (Chap. 35) is clearly indicated in presence of the contra-indications listed above. In selected complex DORV with two viable ventricles, the optimal option is anatomical repair [13, 21, 22, 30]. Nevertheless, a Fontan procedure is still advocated by many centers because of the short term security offered by a Fontan undertaken on two ventricles. However, there are specific issues with the Fontan procedure in complex DORV.

In DORV nc VSD, the VSD is already small in 60 % and can become severely restricted with time. This dramatic complication of the Fontan procedure was described by the Boston group in eight patients in the follow up of Fontan undertaken in DORV nc VSD [25]. The VSD was totally closed in five patients and severely restrictive in three, with supra-systemic LV and compression of the right ventricle. One patient presented with an LV aneurysm. Attempts at surgical VSD enlargement or catheter-based procedures have resulted in almost constant recurrence. This recently reported complication is in favor of also performing a VSD enlargement at the time of the Fontan completion in complex DORV [17] (Chap. 35).

In DORV-AVSD-PS-heterotaxy, the association with a TAPVR is at risk for late pulmonary veins obstruction and pulmonary hypertension. A bi-ventricular circulation would be more suitable (see Chap. 24).

Heart transplantation may be contemplated in rare circumstances.

Outcomes

Mortality

The mortality reported in the literature depends on the anatomic types. In simple forms it is <5 % in DORV-VSD, 5–7 % in DORV Fallot [11] and 0–7 % in Taussig-Bing treated by arterial switch [18, 40].

The mortality is higher in complex forms. In DORV-nc-VSD, there are limited series as shown on Table 23.1. The average mortality, depending on series published, varies from 5 to 15 %. The recent introduction of the tunnelization to the pulmonary artery followed by arterial switch [11, 12, 38] is associated with a mortality of 6 % [13]. Multiple VSD is associated with significant higher mortality [13, 22].

In DORV-AVSD-PS-Heterotaxy, the mortality reported in recent publications is 5 % [13, 30].

Follow Up and Reoperation

The long term follow up in simple forms is equivalent to the surgical repair of respectively: VSD, Fallot and Taussig Bing. In complex forms, the midterm survival in is around 80–85 %. A recent large series from Fu Wai Hospital, Beijing [38] with 380 bi-ventricular repair of DORV, reports that in 67 DORV nc VSD (Table 23.1) the operative mortality was 10 % and the 5 years survival 85 %. Additional information on long term outcome is required.

Late subaortic obstruction at the level of the baffle tunnel and/or the VSD is the most frequent cause of reoperation [13, 38, 41–43] and varies from 5 to 35 % according to the series published. This complication was observed early in our experience and can be avoided by an appropriate enlargement of the VSD at the time of the repair [17, 42]. It is significantly more frequent in DORV nc VSD repair [38]. Intra-operative AV blocks have become seldom [17]. Late ventricular arrhythmias [22], pulmonary valve regurgitation, valved conduit failure, as well as aortic dilation require a life-long surveillance by paediatric and adult congenital cardiologists.

Table 23.1 Biventricular repair of DORV nc VSD

Sakata et al. [10]	Paris	Not given
S Kleinert, R Mee, et al. *JTCVS* (1997) [22]	Melbourne	12
E Belli, F Lacour-Gayet et al. [21]	Paris	23
M Barbero Marcial, et al. *JTCVS* (2000) [24]	Sao Paulo	17
J Artrip, F Lacour-Gayet et al. [13]	Denver	11
Li et al. [38]	Beijing	67

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