CHAPTER 3. SURGICAL OPTIONS

The surgical approach to double outlet right ventricle (DORV) varies according to the given anatomic features [1-5]. The ideal surgical repair is a biventricular repair by connecting the morphologically left ventricle to the aorta and the morphologically right ventricle to the pulmonary artery. There are occasions when the left ventricle can only be connected to the pulmonary artery and the repair is completed by an arterial or atrial switch operation. A biventricular repair is feasible in the majority of patients with two well-developed ventricles, while severe hypoplasia of a ventricle, significant mitral or tricuspid valve pathology, a remote location of the ventricular septal defect (VSD), multiple VSDs and complex form of pulmonary atresia may necessitate a univentricular repair or other type of palliative procedure.

Although well-known pathological classification of DORV based on the relationship between the VSD and the great arterial trunks is useful, the commitment of the VSD to an arterial valve or valves does not always predict the surgical approach [1-3]. Recent classification named by the STS-EACTS [International Nomenclature named after the Society of Thoracic Surgeons (STC) and the European Association of Cardiothoracic Surgery (EACTS)] and adopted by the Association of European Pediatric Cardiology (AEPC) defines four types of DORV based on the clinical presentation and treatment [1, 6-9]: 1) VSD-type, 2) tetralogy-type, 3) transposition of the great arteries (TGA)-type, and 4) non-committed VSD-type (Table 3-1).

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<th>Table 3-1. STS-EACTS-AEPC classification of VSDs in DORV.</th>
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<td>- VSD-type (25%)</td>
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<td>- Tetralogy-type (35%)</td>
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<td>- TGA-type (20%)</td>
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<td>- Non-committed VSD-type (20%)</td>
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* STC, Society of Thoracic Surgeons; EACTS, European Association of Cardiothoracic Surgery; AEPC, Association of European Pediatric Cardiology

1. DORV, VSD-type

This type is characterized by a subaortic or doubly committed VSD without pulmonary stenosis showing the clinical signs of overcirculation from an unrestricted VSD. Usually, it requires a one-stage complete repair within the first 6 months of life. Rarely, in patients with refractory congestive heart failure, initial palliation with a pulmonary artery band to protect the pulmonary vascular bed is required. The repair consists of intraventricular routing of the VSD to the aortic valve using a tunnel-like patch. When the VSD is restrictive, enlargement of the VSD can be required to avoid left ventricular outflow tract obstruction. The VSD is considered restrictive when its diameter is smaller than the diameter of the aortic valve [10-12]. The restrictive VSD is enlarged by making an incision or by resecting a wedge of the septum in an anterior and superior direction as the atrioventricular conduction axis courses along or is a few millimeters away from the postero-inferior margin of the defect. Occasionally, the outlet septum should be resected to construct a
straight tunnel in order to avoid pulmonary outflow tract obstruction. Repair can usually be achieved through a right atrium but occasionally needs a right ventriculotomy.

2. DORV, Tetralogy-type
This type is characterized by a subaortic or doubly committed VSD with pulmonary outflow tract obstruction showing various degree of cyanosis. It requires a complete repair during the first year of life. The VSD can also be a complete atrioventricular septal defect extending toward the outlet with deviation of the outlet septum encroaching on the pulmonary outflow tract. The repair of Fallot-type of DORV consists of relief of subpulmonary outflow tract obstruction through resection of the outlet septum and obstructing muscle bundles when present, and intraventricular tunneling of the VSD to the aorta. Although pulmonary valvotomy or augmentation with an outflow tract patch can be required, preservation of the native pulmonary valve function or restricted enlargement of the pulmonary valve is considered desirable [13-17]. When the right coronary artery or left anterior descending coronary artery takes an anomalous course across the subpulmonary outflow tract, a valved extracardiac conduit needs to be inserted from the right ventricle to the pulmonary artery. Although an early one-stage surgery is preferred, severe pulmonary arterial hypoplasia may need early palliation with a systemic-to-pulmonary arterial shunt.

3. DORV, TGA-type (Taussig-Bing malformation)
This type is characterized by a subpulmonary VSD and is often associated with a degree of subaortic narrowing and tubular hypoplasia, coarctation, or interruption of the aortic arch. The patients present with severe cyanosis and heart failure in the immediate postnatal period as in complete TGA. If applicable, one-stage operation is required in the first or second week of life. When a one-stage surgery is considered to be associated with a high risk, delayed definitive repair after initial palliation with pulmonary artery banding and correction of the aortic arch obstruction and subaortic stenosis has been suggested [18].

Most cases are not associated with pulmonary stenosis and are amenable to either arterial switch procedure with baffling of the VSD to the neo-aorta or intraventricular repair with construction of a direct tunnel from the VSD to the aorta (Kawashima procedure) [1, 19-25]. The Kawashima’s intraventricular repair is applicable when the tricuspid and pulmonary valves are positioned wide apart, leaving enough space for the unobstructed tunnel [2, 3, 25, 26]. The Kawashima procedure requires resection of the outlet septum to secure the unobstructed left ventricular outflow tract. Both arterial switch and intraventricular repair requires enlargement of the VSD when the size of the defect is less than the size of the aortic valve. The incision for enlargement should be on the anterior rim of the VSD as the atrioventricular conduction axis is on or at a short distance from the posteroinferior rim of the VSD. In general, the cases with the great arteries in an antero-posterior relationship tend to have a shorter tricuspid-pulmonary valve distance and are more suitable for an arterial switch procedure, while those with the great arteries in a side-by-side relationship tend to have a wider tricuspid-pulmonary valve distance and are suitable for intraventricular repair [19, 23]. If achievable, the intraventricular repair is advantageous compared to arterial switch operation because there is no need for reimplantation of the coronary arteries, and the risk of later development of aortic regurgitation is avoided [23]. This is especially the case when an unusual coronary artery anatomy is a risk
factor or the pulmonary valve is not considered adequate to function as a systemic valve, such as in patients with prior pulmonary artery banding.

When pulmonary stenosis precludes an arterial switch operation with a VSD closure or when a standard intraventricular tunnel repair is not applicable, an REV (Réparation à l’Etage ventriculaire) procedure can be performed [28-31]. The REV procedure consists of extensive excision of the outlet septum when present, translocation of the pulmonary artery through the gap in the dissected ascending aorta, tunneling of the VSD to the aortic valve and direct anastomosis of the pulmonary artery to the right ventricle using a pericardial patch or monocuspid dacron patch. The REV procedure is a modification of the Rastelli operation which is often complicated by both left and right ventricular outflow tract obstruction due to the long curved nature of the intraventricular tunnel and the lack of growth potential of the external conduit [32, 33]. The REV procedure provides a straighter and wider tunnel from the left ventricle to the aorta and the growth potential for the right ventricular outflow tract as compared to the original Rastelli procedure [28, 29, 33]. It also allows complete repair in infancy. An external valved conduit between the right ventricle and the pulmonary arterial trunk is used only when the pulmonary arterial trunk cannot be directly anastomosed to the right ventricle.

Recently, there has been revival of the Nikaido procedure which consists of mobilization of the aortic root with its valve from the right ventricle, resection of the outlet septum and excision of the pulmonary valve, implantation of the mobilized aortic root in the pulmonary location, patch closure of the VSD, and reconstruction of the right ventricular outflow tract to the pulmonary artery using a pericardial patch [34]. When the stenotic or hypoplastic pulmonary valve is still usable, the pulmonary root can also be mobilized and translocated to the aortic position and the pulmonary outflow tract is augmented by using a transannular pericardial patch. This procedure is called double-root translocation [35].

4. DORV, non-committed VSD-type

The non-committed location of the VSD is defined as a distance of the VSD from both aortic and pulmonary valve greater than the diameter of the aortic valve [8, 36, 37]. The remoteness of the VSD may preclude biventricular repair and instead requires multiple staged operations toward a Fontan circuit [38-40]. However, biventricular repair is often achievable with a long tunneling of the VSD to the aorta or to the pulmonary artery [8, 36, 37, 41]. When the VSD is tunnelled to the pulmonary artery, the aorta and pulmonary arteries should be translocated by using an arterial switch or Nikaidoh operation or double-root translocation procedure depending on the size and healthiness of the pulmonary valve and the absence or presence of subpulmonary stenosis [35]. The surgical procedure for DORV with remote VSD is usually undertaken through a right ventriculotomy, especially when the VSD involves the apical trabecular septum. In the majority of the cases, the VSD is restrictive and needs to be enlarged anteriorly and superiorly [41]. More than one VSD patch is often required to avoid obstruction of the right ventricular inlet or injury to the tricuspid valve tension apparatus [41]. Because the intraventricular tunnel occupies a substantial space in the right ventricle, the right ventricular outflow tract often needs to be enlarged or reconstructed using a conduit. As the corrective surgery involves complex intraventricular repair requiring a long cross-clamp time, staged operation with a Blalock-Taussig shunt or pulmonary artery banding in early infancy and delayed corrective repair in later infancy is often preferred [8].
When biventricular repair cannot be undertaken, univentricular repair is indicated. In addition to the remoteness of the VSD to an arterial valve, straddling of an atroioventricular valve or valves, severe hypoplasia of one ventricle and multiple VSDs are the indications for univentricular repair. Univentricular repair consists of Blalock-Taussig shunt or pulmonary artery banding in the first few weeks of life, bidirectional cavopulmonary connection at 4-9 months of age and modified Fontan operation at 2-4 years. Although biventricular repair is a preferred option, it should be emphasized that biventricular repair requiring a complex intraventricular procedures is associated with a higher requirement for reintervention than univentricular repair [41, 42, 43].

**DORV with discordant atroioventricular connection**

This type is a variant of congenitally corrected TGA. The clinical presentation may vary according to the relationship of the VSD to the arterial valves and the presence or absence of obstruction of the aortic and pulmonary outflow tracts. Until the late 1980’s, TGA or DORV in the setting of discordant atroioventricular connection had been managed by tunneling the VSD to the pulmonary valve so that the morphologically right ventricle supports the systemic circulation and the morphologically left ventricle supports the pulmonary circulation [44]. With increasing incidence of failure of the systemic right ventricle and development of tricuspid regurgitation, the concept of “anatomical repair” using the left ventricle as the systemic ventricle has been introduced. The “anatomic repair” includes combined Mustard/Senning atrial switch procedure, tunneling of the VSD to the pulmonary valve and arterial switch operations [45-48]. Although the “anatomical repair” appears ideal, its long-term outcome is still debated [49]. When an anatomical repair is opted, an elective anatomic repair beyond 6 months of age is recommended since the intracardiac procedure is complex [50-52]. In neonates with severe cyanosis due to severe pulmonary stenosis or hypoplastic pulmonary arteries, a palliative shunt is placed to allow growth of the cardiovascular structures. In neonates with unobstructed pulmonary outflow tract and heart failure, pulmonary artery banding is required until the optimum age for complete repair.

**DORV in Heterotaxy syndrome**

Heterotaxy syndrome, especially the right isomerism is often associated with DORV. The interventricular communication is usually through an AVSD that almost invariably extend toward the aortic valve [9, 37]. In this particular setting, subpulmonary stenosis due to deviated outlet septum is a rule rather than an exception. Some surgeons regard the cases with an AVSD extending toward the outlet and pulmonary outflow tract stenosis as DORV of tetralogy type [8, 9, 37]. Traditionally univentricular repair has been favored because of the proximity of the atrioventricular valve leaflets and their tension apparatuses to the VSD, as well as the frequent association with total anomalous pulmonary venous connection [52]. A more recent study showed that biventricular repair was feasible in the majority of the cases with a low surgical mortality rate [8, 9]. The intraventricular tunneling of the VSD to the aorta requires division of the anterior bridging leaflet of the common atrioventricular valve. Both biventricular and univentricular repairs are
associated with high risks when there is atrioventricular valve regurgitation or pulmonary venous obstruction and when the patient presents in the early neonatal period [52].

DORV with Abnormal Chordal Insertion or Straddling of the Atrioventricular Valves

When the tricuspid valve has an abnormal chordal attachment to the outlet septum, the outlet septum is not resected, but mobilized with the chordal insertions [3, 29, 54-56]. After the intraventricular tunnel is constructed using a patch, the mobilized outlet septum is sutured to the patch. A similar technique can also be used for those with tricuspid chordal attachment to the subpulmonary and/or subaortic outflow tract [56]. More extensive chordal attachment of the tricuspid or mitral valve all around the VSD margin precludes biventricular repair [56]. When there is straddling of the tricuspid or mitral valve, the abnormal chordae or papillary muscle can be retracted toward the ventricle that the straddling valve belongs to and the VSD is closed on the opposite side of the septum [56].

REFERENCES

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