

# Double outlet right ventricle

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**Creation date:** July 2002

**Updated:** December 2003

**February 2005**

**Scientific Editor:** Professor Robert H Anderson

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## Abstract

*The term "Double Outlet Right Ventricle" (DORV) refers to any cardiac anomaly in which both the aorta and pulmonary trunk originate, predominantly or entirely, from the right ventricle. A wide spectrum of malformations occurs. Some have features similar to Fallot's Tetralogy, others resemble Transposition with ventricular septal defect and many other variations occur, some simple and others complex. The clinical picture varies widely, depending on the combination of abnormalities present. Cyanosis is frequent, but not universal. Heart failure with breathlessness, poor feeding and slow weight gain is also common. The malformation results from an error in formation of the outlet part of the ventricular loop during early embryonic life, probably within the first three to four weeks after conception (between 5 and 6 weeks gestational age). In most cases the defect is "sporadic" in pattern and there is no identifiable genetic cause. The primary diagnostic tool is echocardiography. The overall frequency is not well documented as the condition overlaps with a range of defects and is often not categorized solely as DORV in epidemiological studies of congenital heart disease. In its many forms it is probably present in 3% or more of all congenital heart defects, thus indicating a birth incidence of less than 1:3,000. Most affected patients can undergo surgical repair.*

## Keywords

ventricular septal defect, Taussig-Bing Malformation, Eisenmenger's "Anomaly", Fontan procedure

## Disease name and synonyms

- Double Outlet Right Ventricle (DORV)
- Taussig-Bing Malformation
- Tetralogy of Fallot with extreme dextroposition of the aorta

- Eisenmenger's "Anomaly" (Eisenmenger Ventricular Septal Defect)

## European Paediatric Cardiac Code

Reference of Double Outlet Right Ventricle is 01.01.04.

## Introduction

The term "Double Outlet Right Ventricle" refers to any cardiac anomaly in which both the aorta and pulmonary trunk originate, predominantly or entirely, from the right ventricle (RV) (1). In this situation, the left ventricle (LV) has no direct outlet to either great artery and ejects through an interventricular communication, usually referred to as a "ventricular septal defect" (VSD), (which is almost invariably present) into the right ventricle. Rarely there may be no "VSD" and the LV is then extremely hypoplastic.

The literature on DORV is substantial, and includes descriptions of a large range of anatomic variations (2, 3) which are reflected in many different clinical presentations. The medical and surgical management of this entity is also heterogeneous and needs to be tailored to the particular problems of each affected child (4-7).

A logical and step-by-step approach to the diagnosis and classification of DORV is essential. Atrial arrangement (and venous connections) must be ascertained, and the atrioventricular connections established with certainty. Identification of the way in which the two great arteries arise from the right ventricle is important, including their inter-relationships and the nature and severity of any obstructive problems. The size, site, and morphology of the interventricular communication, which is categorised in a similar way to that used for isolated ventricular septal defects, are also of great importance, particularly in reference to their relationship with the arterial outlets. Attention must also be paid to the anatomy and function of the atrioventricular valves (especially the mitral valve), and to the presence of other associated cardiac defects, which are frequent. Separation of the arterial valves from the atrioventricular valves by a bar of ventricular muscle (e.g. subaortic "conus") is frequent but not universal. Such separation may also be seen in occasional patients with other defects, including subaortic stenosis, VSD and Tetralogy of Fallot without DORV. Thus the presence of a "subaortic conus", once regarded as characteristic of the anomaly (8), is not a useful criterion for diagnosis of DORV.

The categorization of each case should be individualized, just as the medical and surgical management need to be tailored to cater for the particular problems of the individual case. Nonetheless, certain variants of double outlet right ventricle do occur frequently enough to merit separate discussion. These will be outlined below.

### **The commoner variants (1) are:**

1. DORV with subaortic ventricular septal defect (interventricular communication), aorta to right of pulmonary trunk, and pulmonary stenosis (Fallot type), (9).
2. DORV with aorta to right and subpulmonary interventricular communication [VSD] (Taussig Bing Malformation), (10).
3. DORV with subaortic interventricular communication and without pulmonary stenosis (aorta to right of pulmonary trunk), (11, 12). (Eisenmenger anomaly), (13).

### **Less frequent variants include DORV associated with:**

1. With non-committed VSD (interventricular communication), usually with the aorta to the right of the pulmonary trunk (14).
2. With doubly committed interventricular communication (VSD) (12, 15).
3. With subaortic interventricular communication (VSD) with the aorta to the left of the pulmonary trunk along with pulmonary stenosis (16).
4. With discordant atrioventricular connection (17), (Aorta usually to the left of the pulmonary trunk).
5. With mirror-image atrial arrangement (1), (Any of the above-mentioned variations may occur).
6. With isomeric atrial appendages and, hence, ambiguous atrioventricular connection (18-20).

### **Differential diagnosis**

The malformation "Double Outlet Right Ventricle" is not a single diagnostic entity. It may occur as part of a range of cardiac defects – often complex. Because the clinical picture is extremely variable (1), it may present with features similar to such conditions as:

- [Fallot's Tetralogy](#)
- [Transposition of the Great Arteries](#)
- [Coarctation of the aorta](#)
- [Isolated Ventricular Septal Defect](#)

DORV is very frequently present in infants with complex cyanotic heart disease, such as occurs in the presence of Isomerism of Right Atrial Appendages (sometimes called "Asplenia Syndrome") (18). Such infants often present with severe cyanosis and / or respiratory distress in the early newborn period and the differential diagnosis includes many of the defects that can lead to severe symptoms in the early neonatal period, including pulmonary atresia, transposition, critical pulmonary stenosis, Ebstein's anomaly and many complex congenital cardiac defects.

Establishment of the diagnosis depends on accurate imaging of the intracardiac anatomy. In the past this usually involved cardiac catheterisation and angiography. Currently

diagnosis is usually made with echocardiography.

### Frequency

The condition was present in approximately 3% of infants with severe cardiac malformations who died in early infancy (21). The overall frequency is not well documented as the condition overlaps with a range of defects and is often not categorized solely as "Double Outlet Right Ventricle" in epidemiological studies of congenital heart disease. In its many forms it is probably present in 3% or more of all congenital heart defects – indicating a birth incidence of less than 1:3,000.

### Clinical description

#### ***Double outlet right ventricle with subaortic interventricular communication (VSD) and pulmonary stenosis (Fallot type), (22)***

In this common variant, the clinical picture is indistinguishable from classic Tetralogy of Fallot. A systolic murmur is usually heard from the newborn period (due to the pulmonary stenosis). Cyanosis develops and progresses during the early months of life. Hypoxic spells may occur. Management is largely surgical and complete repair is performed as a primary procedure if feasible (usually within the first six to twelve months). It involves placement of an intraventricular patch to direct left ventricular blood to the aorta and relief of right ventricular outflow obstruction (pulmonary stenosis). Some infants, especially those who develop severe cyanosis early in infancy, may have a preliminary "shunt" operation prior to complete repair.

#### ***Double outlet right ventricle with subpulmonary interventricular communication (VSD) (Taussig Bing Malformation), (10,23)***

The clinical picture is essentially the same as that of Transposition with associated VSD. Presentation is often one of cyanosis in the newborn period with early development of symptoms of "heart failure". Increasing breathlessness, poor feeding and slow weight gain are prominent features. A murmur is usually present, even in the absence of pulmonary stenosis (which is uncommon). An associated coarctation of the aorta is frequent and usually leads to very early onset of heart failure (e.g. in the first week of life).

Management is largely surgical and requires a repair that leaves the left ventricle connected to the aorta and the right ventricle feeding the

pulmonary circulation. This may be achieved by the use of an intraventricular patch connecting the LV to the aorta (24, 25) (which is often difficult to achieve). Alternatively a patch may be placed to connect the LV to the pulmonary artery (which is easier to perform) and an "Arterial Switch" operation (as for transposition) may be done so that the left ventricle is connected to the aorta and the RV to the pulmonary circulation (7, 26, 27). The operation is usually carried out within the first 1 – 3 months.

#### ***Double outlet right ventricle with subaortic interventricular communication and without pulmonary stenosis (28)***

The blood from the left ventricle is directed preferentially to the aorta (as the interventricular communication is close to this artery). In the absence of any pulmonary stenosis there is increased flow to the pulmonary circulation and the clinical picture resembles that of a large, isolated, VSD. Cyanosis is very mild or absent and symptoms usually develop gradually over the first few months of life, with poor feeding and weight gain accompanied by increasingly apparent breathlessness and an increasingly loud heart murmur, which may not be present initially. If an associated coarctation is present symptoms appear earlier and are more severe. Surgical repair is usually performed as a primary procedure at an age between one month and six months. Repair involves placement of an intraventricular patch to direct left ventricular blood to the aorta.

### Other variants

In other malformations in which DORV is present the clinical picture commonly resembles that seen in one of the variants described above. The time of onset and severity of symptoms tend to depend often on associated defects, such as coarctation, which usually leads to early onset of heart failure or pulmonary stenosis / atresia, associated with cyanosis as the main feature. Most affected patients can undergo surgical repair (29, 30), though in the more complex variants complete "repair" may not be feasible (especially those with Isomerism of Right Atrial Appendages) and palliation involving a Cavo-Pulmonary shunt or Fontan operation may be performed (30).

### Etiology

The malformation results from an error in formation of the outlet part of the ventricular loop during early embryonic life – probably within the first three to four weeks after conception (5-6 weeks gestational age), (31, 32). In most cases

the defect is 'sporadic' in pattern and there is no identifiable genetic cause. A small number of familial cases have been reported and the defect has been produced in animal models by deletion of specific genes (33, 34) – especially those involved with migration of neural tube cells into the branchial arch area and the developing heart (33). A chromosomal micro-deletion in humans (22 q11 deletion) is associated with a range of cardiac abnormalities including, in some cases, forms of DORV (35, 36).

### Diagnostic methods

As indicated above the primary diagnostic tool is echocardiography (37, 38). This requires high-quality 2D imaging, with high frequency (e.g. 10 MHz) transducer for infants, and facilities for Doppler interrogation and colour flow mapping. Cardiac catheterisation and angiography may provide additional information in selected cases, but are not usually essential in making the initial diagnosis. Simple tests such as ECG and Chest X-ray have their place and provide some helpful information, but the findings are not specific and are of limited diagnostic utility.

### Prognosis

Complete surgical repair (Biventricular repair) is achievable in most patients with DORV, with low risk (< 5% ) (29, 30). In complex DORV (patients with abnormal atrial arrangement, discordant atrioventricular (AV) Connections, common atrioventricular valve, hypoplasia of one ventricle, straddling AV valves, multiple VSDs or Pulmonary Atresia) the risk for complete repair is higher (e.g. equal or superior to 20%). For these complex patients a Fontan procedure, if they are suitable for this type of surgery, is associated with lower operative mortality (30). Specific risk factors that indicate greater hazard for biventricular repair include coexisting aortic arch obstruction (Coarctation or Arch Interruption), the presence of multiple VSDs and age less than 1 month at the time of repair (30).

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