

Double-Outlet Right Ventricle

Patient Information Series – What you should know, what you should ask.

What is double-outlet right ventricle (DORV)?

The heart is made of 4 chambers and 4 valves which is divided into right and left sides. The lower chambers of the heart are called ventricles. In a normal heart, the right ventricle pumps low oxygen (blue) blood returning from the body into the lungs through the pulmonary artery. The left ventricle pumps high oxygen (red) blood from the lungs into the body through the aorta.

DORV is a rare cardiac abnormality, where *both* the pulmonary artery and aorta arise from the right ventricle of the heart. There is usually a defect or hole in the wall between the left and right ventricles (ventricular septal defect/VSD). There are different types of DORV. There may be narrowing of the one of the arteries leaving the heart or the arteries may be in an unusual position relative to each other. DORV is a heart defect that needs open heart surgery to correct it.

How does DORV happen?

During the early development of the fetus, the heart is formed by twisting and turning itself in a rightward direction. Sometimes problems arise during this process, resulting in the arteries coming off the heart in abnormal locations.

What are the possible causes?

DORV is often an abnormality that arises without a clear cause. There can be genetic causes of DORV and sometimes there can be a genetic syndrome associated. Poorly controlled diabetes mellitus in early pregnancy can be associated with an increased chance of DORV.

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What additional tests or workup may be needed?

- A fetal echocardiogram will be performed to confirm the diagnosis and give information about what treatment will be needed
- A detailed fetal anatomy ultrasound scan should be done to assess for any other fetal abnormalities
- A consultation with Genetics to discuss the option of prenatal genetic testing
- A consultation with Pediatric Cardiologist is advised to help monitor the pregnancy, counsel parents about options for management and prepare for the birth of a baby that will need special cardiac attention after birth.

What are the things to watch for during pregnancy?

If DORV is confirmed, you will likely be followed by a high-risk obstetrician with close monitoring of fetal growth and well-being by ultrasound.

What should be expected after delivery?

Delivery will usually occur at a centre where pediatric cardiologists and cardiac surgeons are available nearby. Whether the baby needs urgent medication or procedures after delivery depends on the type of DORV and will be explained to you by your doctor. Many babies will require a period of treatment in the NICU. Some babies will need an intravenous medication called Prostaglandin to maintain their lung or body blood flow while they await surgery. The timing of surgery can range from within the first week of life up to 6 months or more of age, depending on the exact type of DORV.

Will it happen again?

- Most cases of DORV are sporadic and not inherited.
- For isolated DORV, the risk of recurrence is about 1.5 to 2 in 100.
- If a chromosomal problem is present (e.g., trisomy 18), the recurrence risk is very low.

How can I prevent this?

If you have Diabetes Mellitus, your blood sugar should be well controlled before planning to get pregnant.

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