Double Outlet Right Ventricle

A GUIDE TO HELP UNDERSTAND YOUR BABY'S HEART

PROVIDED BY THE
CENTER *for* ADVANCED FETAL CARE
the FETAL HEART PROGRAM

UNIVERSITY *of* MARYLAND
MEDICAL CENTER

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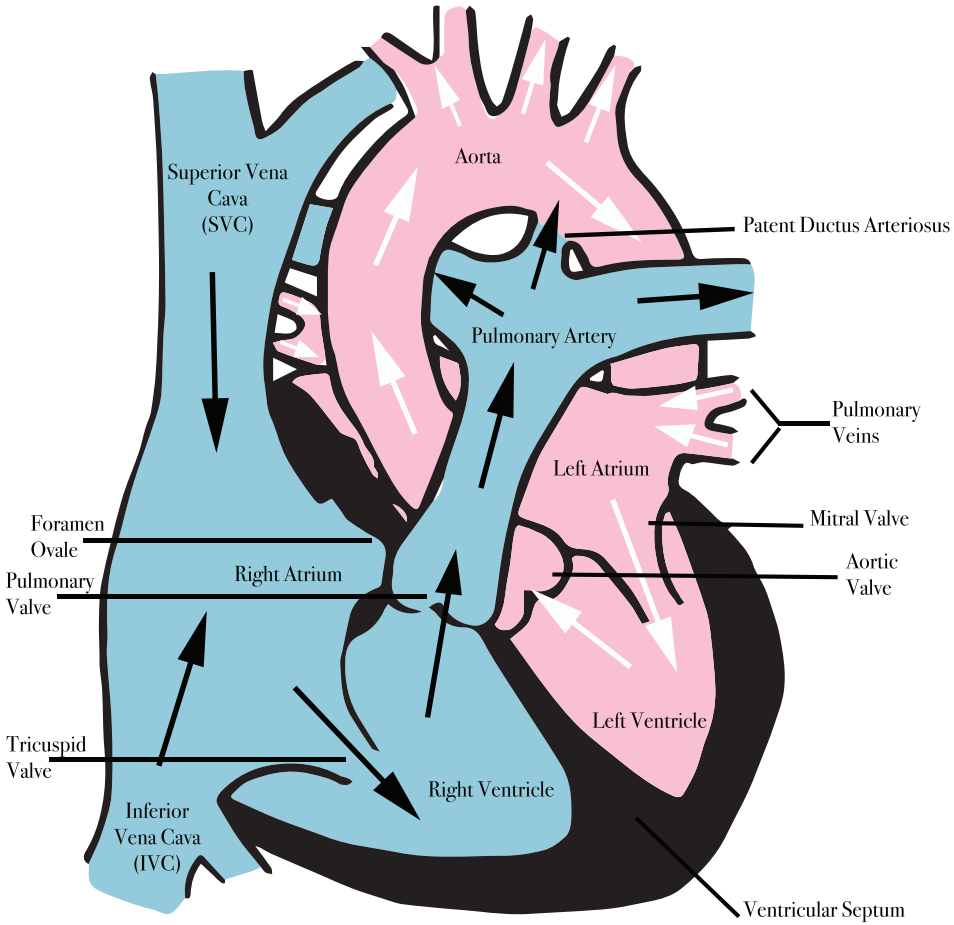
This guide will help you understand your child's heart. It is not a diagnosis and should never be used instead of medical advice.

Our goal at the Center for Advanced Fetal Care is to assist you on the journey ahead and help educate you to better communicate with your team of physicians, friends and family.

Illustrations by Dr. Alicia Chaves

The Normal Heart



The heart is a complex organ which pumps blood through the body. It drives the circulatory system, which carries oxygen and nutrients to the vital organs through a system of arteries and veins.

The heart has four chambers. The top two chambers are called the atria, which are separated by the atrial septum. The bottom two chambers are the ventricles, which are separated by the ventricular septum. As blood passes through each individual chamber, it exits through a valve. Each side of the heart works as its own pump.

Pump 1 - Right side pumps blood to the lungs. (blue on diagram) Blood travels from the right atrium through the tricuspid valve into the right ventricle. From the right ventricle, it travels through the pulmonary valve into the pulmonary artery to the lungs. Oxygenated blood from the lungs returns to the left atrium through the pulmonary veins.

Pump 2 - Left side pumps blood to the body. (pink on diagram) From the left atrium, oxygenated blood travels through the mitral valve to the left ventricle. Then, blood exits through the aortic valve to the aorta, the main artery, sending blood to the rest of the body. Once blood has supplied oxygen to the vital organs, it returns to the right atrium through the inferior vena cava (IVC) and the superior vena cava (SVC), the main veins of the body, to begin the process again.

What is a congenital heart defect?

A congenital heart defect is an abnormality of the structure and/or function of the heart. The defect typically develops during the early stages of pregnancy.

Why did this happen to my baby?

A congenital heart defect is the most common abnormality found in babies. Congenital heart disease may occur due to environmental factors, chromosome abnormalities, or genetic conditions; the majority of heart defects are multifactorial, which means that it occurs because of interactions between genes, chance, and the environment. In most cases, there is not an explanation for why a baby is born with a heart defect.

It is important to remember that any baby can have a congenital heart defect. Neither parent is to blame. Our hospital offers monthly support groups for families with children born with a congenital heart defect.

For more information, visit the fetal heart program online at: umm.edu/programs/fetalheart/patient-information/resources

What is Double Outlet Right Ventricle?

Double outlet right ventricle (DORV) is a congenital heart defect in which the aorta (the blood vessel that takes blood to the body) and main pulmonary artery (the blood vessel that takes blood to the lungs) both come from the right ventricle of the heart, rather than the normal arrangement explained previously.

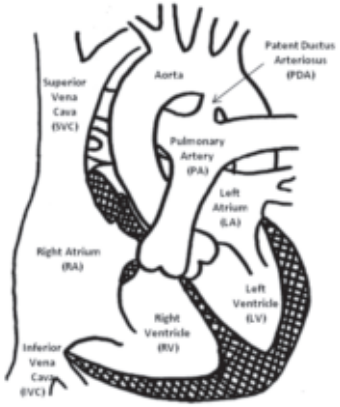
There are usually other cardiac defects that occur with DORV. The most common defects are a ventricular septal defect (VSD), pulmonary stenosis, and/or aortic stenosis, which are explained below. Since each case is slightly different, your doctor will help to clarify the specifics of your baby's condition to make an individual plan for your family.

Ventricular Septal Defect (VSD): A hole in the heart between the two ventricles. The location and size of the VSD will determine how blood will flow through the heart.

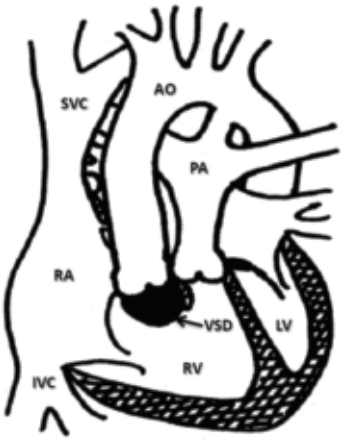
Pulmonary Stenosis: Narrowing of the pulmonary artery which can result in decreased blood flow to the lungs and low oxygen levels. If oxygen levels become too low, the baby's skin could begin to appear blue in color.

Aortic Stenosis: Narrowing of the aorta which can result in decreased blood flow to the body. Most often no signs or symptoms are seen in a newborn with aortic stenosis.

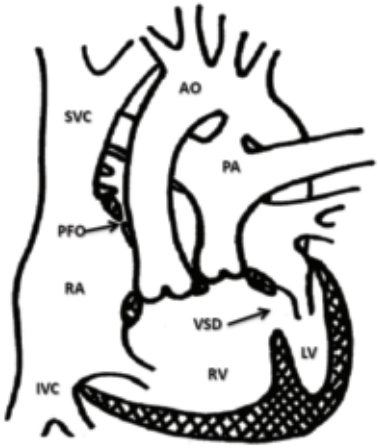
Normal Heart



Double Outlet Right Ventricle with subaortic VSD



Double Outlet Right Ventricle with subpulmonary VSD



For more information visit the fetal heart program online at:
umm.edu/programs/fetalheart/health-information/services/double-outlet-right-ventricle

How is Double Outlet Right Ventricle Diagnosed?

Double outlet right ventricle is usually diagnosed during a detailed ultrasound of the fetal heart called a fetal echocardiogram. This can be done at any point in the pregnancy that an abnormality is suspected.

Our goal at the University of Maryland is to diagnose fetal heart defects as early as possible. We strive to assess the heart during the first trimester screening between 12-14 weeks and perform an early echo if a heart problem is suspected, or if the pregnancy is at an increased risk to have a heart defect. Since the heart is very small at this early age, our ability to assess and diagnose a heart condition, like DORV, depends on the fetal position and the nature of the abnormality.

If a first trimester screen was not performed, or a problem was not detected because of the limitations of the ultrasound, it is often detected at the second trimester ultrasound. In general, a targeted anatomy ultrasound is performed between 18-20 weeks to assess all of fetal structures, including the heart.

If a heart defect is suspected in the first or second trimester, a fetal echocardiogram is done between 22-24 weeks to confirm or clarify the suspected diagnosis.
Occasionally, DORV may not be diagnosed until the baby is born. After birth, symptoms such as a bluish skin tint or a heart murmur (sound made by abnormal blood flow) can indicate the problem.

In any of these cases, an echocardiogram after the baby is born confirms and gives detail about the specific condition of the DORV.

What causes Double Outlet Right Ventricle?

Doctors are often never able to figure out why a baby developed DORV. When a baby has DORV but no other birth defects, it is usually "multifactorial", meaning that a lot of different genetic and nongenetic factors combined to cause it. Sometimes environmental factors like maternal diabetes or certain medications taken by a mother during pregnancy can cause DORV. When DORV is seen in combination with other birth defects, it is more likely that the baby could have a genetic disorder or a chromosomal condition associated with medical and/or developmental issues in addition to the DORV. During pregnancy, either chorionic villus sampling (CVS) or amniocentesis can test for many chromosomal conditions. These procedures are associated with a small risk of miscarriage, however many providers and patients believe that the benefits outweigh the risks. If a family chooses not to have these tests during pregnancy, all newborn babies with DORV will be checked for genetic and chromosomal conditions.

What can I expect during my pregnancy?

If your baby is diagnosed before birth with double outlet right ventricle, a team of specialists will care for you and your unborn baby. The team at the University of Maryland Medical Center includes a maternal fetal medicine specialist (MFM), cardiologists (fetal and pediatric), genetic counselor, neonatologist, and a pediatric cardiac surgeon. Your baby will be monitored closely by fetal ultrasounds and a delivery plan will be discussed among you, your obstetrician, and the various other specialists. Induction of labor may be scheduled for a pregnancy affected with DORV to ensure that your care team is present at delivery. If there are no maternal or fetal issues other than the baby's DORV, our goal is for you to have a normal delivery, with no intervention.

What can I expect after my baby is born?

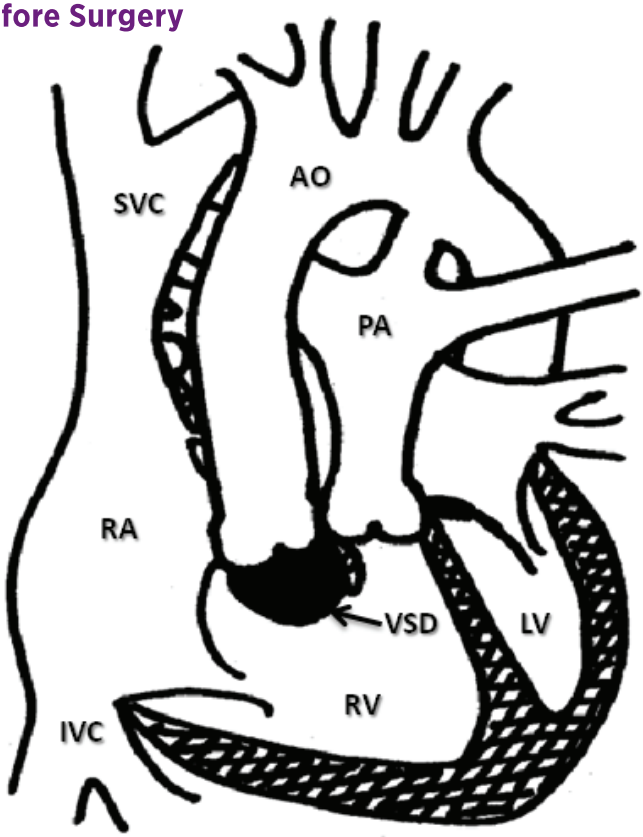
After delivery, your baby will be taken to the neonatal intensive care unit (NICU). The specialists in the NICU will closely monitor your baby's breathing and oxygen levels. An echocardiogram will be performed shortly after birth. If your baby has normal oxygen levels, he or she may be able to go home soon after birth with frequent monitoring as an outpatient. Surgery will be needed in the future to fix the heart defects.

How is Double Outlet Right Ventricle treated?

The ultimate goal of surgery is to divide the heart into two separate ventricles, with the aorta coming from the left ventricle and the pulmonary artery coming from the right ventricle. The treatment plan for a baby born with DORV depends on the arrangement of the vessels, the location of the VSD (hole between the two ventricles), and if there is pulmonary and/or aortic stenosis (narrowing of the vessel).

The most common location for the VSD is near the aorta. In this case, the surgeon sews a patch from the ventricular septum to the aorta, which closes the VSD. This allows blood from the left ventricle to exit correctly through the aorta. In these cases, pulmonary stenosis is often present, which the surgeon will also repair.

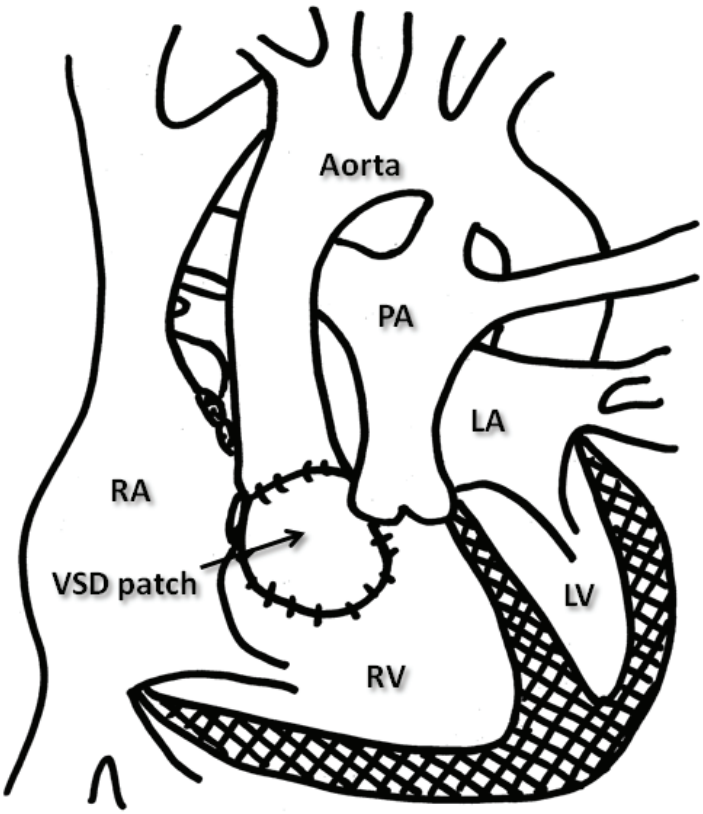
Before Surgery



If the VSD is near the pulmonary artery (sometimes called a Taussig-Bing anomaly) a more complex heart surgery is required. In order to obtain the goal of dividing the heart into two ventricles with the aorta coming from the left and the pulmonary artery coming from the right, multiple steps are needed. The arteries will need to be switched and the VSD must be closed so that the left ventricle is connected to the new aorta. In these cases, aortic stenosis is often present, which may require further surgery.

If the VSD is not near either vessel, the surgeon may not be able to divide the heart into two ventricles. In these cases, other surgeries would be done to create a single pumping chamber, supplying blood to both the pulmonary artery and the aorta.

After Surgery



Will my baby have a normal childhood?

In general, the long term prognosis for DORV is good. Your child will need lifelong follow up visits with a cardiologist. Some patients may need an additional surgery or procedure. These procedures could consist of a cardiac catheterization to evaluate the repair or fix areas that may narrow. A patient with DORV may be on medications. Most children born with DORV are allowed to participate in sports, with generally few restrictions, if any. Some patients may develop an arrhythmia (abnormal heartbeat) later in life that may require medications or a pacemaker. Your cardiologist will decide if your child should avoid any activities.

Will we have another child with a congenital heart defect?

Studies suggest that if you have one child with a congenital heart defect, your risk of having another child with a heart defect is about 2%-3%. If your baby's congenital heart defect is associated with a chromosome abnormality or a genetic syndrome, a genetic counselor can discuss with you and your family the risk of having another baby with the same condition. In any future pregnancies, we highly recommend nuchal translucency screening with an early fetal echocardiogram during your first trimester. Then, we recommend a targeted anatomy ultrasound between 18-20 weeks, and a fetal echocardiogram between 22-24 weeks.

How can the Fetal Heart Program help?

The Fetal Heart Program at the University of Maryland Medical Center is dedicated to the care and support of you and your unborn child. Our world class program aims to diagnose congenital heart defects as early, and as accurately as possible. We strive to create personalized prenatal care and optimize your delivery plan. Our multidisciplinary team is devoted to you and your baby's needs before and after birth.

It has been
our privilege
to care for you and your child.

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