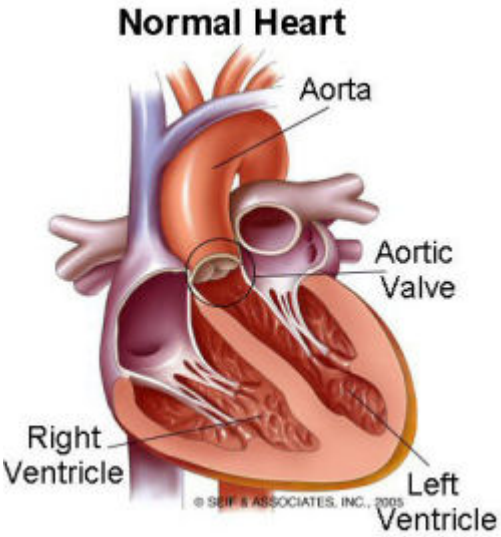
Double Outlet Right Ventricle

Double Outlet Right Ventricle (DORV) means there is a heart problem. It is a heart condition that babies are born with. There are many different types of DORV. Some babies are critically ill from the moment they are born, and some have so few symptoms it may be hard to tell they have a problem.



The normal heart is divided into four parts:

- Upper.
- Lower.
- Right.
- Left.

The two upper chambers of the heart, called the right atrium and left atrium, receive blood as it comes back to the heart. The blood is then sent to the lower chambers, the ventricles. The two bottom chambers, called the right ventricle and left ventricle, pump the blood out of the heart.

The right ventricle pumps blood to the lungs, and the left ventricle pumps blood to the body. The blood leaves the ventricles by two different blood vessels. In DORV, both of these blood vessels come from the same pumping chamber, the right ventricle.

How DORV affects your baby depends on the other problems in the heart. These other problems may include:

- Holes in the walls that divide the right and left sides of the heart.
- The blood vessels may be too small, or may be missing valves.
- There may be a narrowing in the aorta (*coarctation*).
- Your child may only have one ventricle instead of two.

Every baby is unique. You should talk to your baby's cardiologist so that you can understand what other conditions your baby's heart has and how the heart can be fixed, and which problems will need surgery.

CAUSES

DORV occurs at about the 7th week in the womb. There are some genetic conditions that are associated with DORV, but many times no cause can be found. Your baby should be tested for genetic causes, and you should ask the cardiologist if the baby has a syndrome (multiple symptoms due to a genetic cause).

SYMPTOMS

Because the spectrum of DORV is so broad, it is difficult to say how your baby will act. In general, DORV falls into two broad categories:

- Cyanotic (blue), with the symptoms of:
  - Blue color in lips or fingertips (because of less oxygen in the blood).
  - Rounded fingertips and toes.
  - Worsening blueness when crying.
- Acyanotic (pink) with the symptoms of:
  - Pink color (because of too much blood with oxygen).
  - Fast and/or hard breathing.
  - Fatigue and/or sweating when eating.
  - Sleeping more because of easy fatigue.

Both cyanotic and acyanotic babies may have difficulty gaining weight. These babies may need special formula

or feeding tubes to help them eat enough. Discuss your baby's individual condition with your cardiologist so that you can know whether your baby is doing well with their specific problem.

DIAGNOSIS

To determine whether a baby has DORV, the doctors will do most, if not all, of the following tests:

- Examine your baby – to check oxygen level in the blood, blood pressure, listen to the heart and lungs, and check for other possible findings including evidence of a syndrome.
- Chest X ray – this will check the baby's heart size and shape, and show how much blood is flowing to the lungs.
- EKG – this checks the heart's electric circuit and looks for disturbances in heart rhythm (*arrhythmia*).
- Echocardiogram – this is an ultrasound of the heart that will show all of the heart and how the blood flows in the heart. There is no radiation in an echocardiogram.
- Cardiac catheterization – a cardiologist may use a catheter (a long IV that can go inside of the heart) to check oxygen level and blood pressure in all the parts of the heart. The baby will be asleep during this procedure. A cardiac catheterization does have radiation, as this is continuous X-ray.

TREATMENT

All babies with DORV require heart surgery. Because there are many different types of DORV, there are many different surgeries that can be done.

Until the time of surgery, your baby may be on medicines to help the heart. These medicines include:

- Diuretics – these help the body lose excess fluid that may be building up in the lungs. The baby will urinate more if he/she is taking a diuretic.
- Blood pressure medicine – this will lower the baby's blood pressure to help blood flow in a more useful way in the heart.
- Digoxin – this helps the heart pump and can also treat some heart rhythm problems.
- Prostaglandin – if your baby's lungs are not receiving enough blood, this medicine will be used to open up an extra blood vessel (PDA) to help. This is an IV medicine that needs to be given continuously and always in an ICU.

The surgery your baby will have depends on the anatomy of the heart. The surgeon and cardiologist must explain the surgery to you in detail before the baby can go to the operating room. Some babies simply require a patch to close a hole between the two ventricles. Some babies require multiple surgeries throughout infancy and childhood. Every baby is unique.

PROGNOSIS

Because of the variability in the anatomy, the prognosis is variable as well. Most often, your child will need to take medicines throughout their life. If the baby has a syndrome, the prognosis may be different than a baby without a syndrome. As advances in medicine and surgery emerge, new treatment options become available. Be sure to keep open communication with your baby's cardiologist. The more you understand about your baby's heart, the better.

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