CONDITION

Double outlet right ventricle

Double outlet right ventricle is a congenital heart condition in which both main arteries connect mainly to the right ventricle.

WHAT IT IS
Congenital difference in the heart's main artery connections

BODY SYSTEM
Heart and circulation

MAIN SYMPTOMS
Blue or grey skin, fast breathing, poor feeding, slow growth

WHAT CAUSES IT
Heart development difference before birth, usually unknown

HOW IT IS DIAGNOSED
Fetal or postnatal echocardiogram

MAIN TREATMENT
Heart surgery and lifelong cardiology follow-up

CAN IT BE CURED?
No - surgery repairs circulation; lifelong monitoring remains

IS IT AN EMERGENCY?
Sometimes - severe blueness or breathing trouble

LONG-TERM OUTLOOK
Many survive into adulthood; later heart problems may occur

Overview

Double outlet right ventricle, or DORV, is a heart condition present from birth. It develops while the heart is forming during pregnancy.

Normally, the pulmonary artery leaves the right ventricle and carries blood to the lungs, while the aorta leaves the left ventricle and carries oxygen-rich blood to the body. In DORV, both arteries connect entirely or mainly to the right ventricle.

There is also usually a hole between the ventricles, called a ventricular septal defect, or VSD. Blood from the left ventricle must pass through this hole to reach a main artery.

DORV includes several heart arrangements. Its effects depend on:

Some arrangements send too much blood to the lungs and cause heart failure symptoms.

Others send too little, causing low oxygen levels. Most people need surgery planned around their exact heart structure.

- The VSD's position relative to the arteries
- Whether blood flow to the lungs is narrowed or blocked
- The positions of the aorta and pulmonary artery
- Other heart differences

Symptoms

Symptoms may begin during the first days, weeks, or months after birth. Possible symptoms include:

Blue or grey colouring is called cyanosis. On darker skin, it may be easier to see on the lips, tongue, gums, palms, soles, or around the eyes.

When too much blood reaches the lungs, a baby may develop heart failure. This means the heart cannot meet the body's needs; it does not mean the heart has stopped.

After childhood repair, some people have few symptoms. Others later develop reduced exercise tolerance, palpitations, breathlessness, or swelling.

- Blue, grey, or unusually pale lips, tongue, or skin
- Fast, difficult, or noisy breathing
- Sweating or tiring during feeds
- Long feeds or drinking less than usual
- Poor weight gain or slow growth
- Unusual sleepiness or reduced alertness
- A fast heartbeat
- Swelling

Causes and risk factors

DORV occurs when the heart and arteries do not form in the usual way early in pregnancy. Most cases have no identifiable cause, and the condition is not caused by something a parent did.

DORV sometimes occurs with a chromosome or gene difference or as part of a broader

pattern of congenital conditions. However, many children have no identified genetic condition.

A family history of congenital heart conditions may increase the chance of a heart difference, but DORV is not usually inherited predictably. Genetic counselling can explain testing and the possible chance of recurrence in a future pregnancy.

Diagnosis

DORV may be suspected during a pregnancy ultrasound. A fetal echocardiogram can examine the baby's heart chambers, valves, arteries, and blood flow in more detail.

After birth, possible signs include low oxygen readings, a heart murmur, breathing problems, poor feeding, or blue or grey colouring. An echocardiogram usually confirms the diagnosis and shows how blood moves through the heart.

Other tests may help define the anatomy or prepare for surgery:

Genetic testing may be offered when there are other congenital differences, developmental concerns, or signs of a chromosome or gene condition.

- Pulse oximetry to measure blood oxygen
- An electrocardiogram to record heart rhythm
- A chest X-ray to assess the heart and lungs
- Cardiac MRI or CT for detailed images
- Cardiac catheterisation to measure pressures and map blood flow

Treatment and management

A specialist congenital heart team plans treatment based on the VSD's location, artery positions, blood flow to the lungs, ventricular function, and other heart differences.

A newborn with dangerously low oxygen or unstable circulation may need intensive care, breathing support, and medicines that maintain blood flow until a procedure or surgery is possible. Other medicines can reduce fluid buildup or help manage excess blood flow, but they do not correct the artery connections.

Most babies need surgery. A surgeon may create a pathway directing blood from the left ventricle through the VSD to the aorta. Surgery may also repair narrowing or close other openings.

Depending on the anatomy, treatment may involve moving a main artery, placing a tube called

a conduit between a ventricle and the pulmonary artery, or performing operations in stages. If the heart cannot support two separate pumping systems, single-ventricle or Fontan-type operations may direct returning blood to the lungs without passing through a ventricle.

These operations are not interchangeable. Imaging and surgical assessment determine the appropriate approach.

Ongoing care may include:

Good dental care lowers the risk of infective endocarditis, an infection of the heart's inner lining or valves. Antibiotics like amoxicillin or doxycycline before dental procedures are recommended only for certain conditions and stages of repair. The congenital heart team can advise whether they are needed.

- Regular congenital cardiology visits
- Echocardiograms and heart rhythm tests
- Monitoring of growth, feeding, learning, and development
- Medicines for heart function or rhythm
- Further catheter procedures or surgery

Complications

Without effective treatment, DORV can cause low oxygen, poor growth, heart failure, lung blood vessel damage, or life-threatening circulation problems.

Possible complications after surgery include:

Some children have developmental, learning, attention, or coordination difficulties. Developmental assessment and early support may help. Further catheter procedures or operations may be needed as the child grows or repaired areas change.

- Narrowing or blockage in a surgically created pathway
- Deterioration of a conduit
- Leaking or narrowed valves
- Reduced pumping strength
- Abnormal heart rhythms
- Restricted blood flow
- Infective endocarditis

- Reduced exercise capacity

Prevention

DORV usually cannot be prevented, and most cases have no known cause.

Before pregnancy, people can review health conditions and medicines with a healthcare professional, particularly if they have diabetes or take medicines that may affect pregnancy. These steps support a healthy pregnancy but cannot guarantee prevention.

If congenital heart disease has occurred in a family, genetic counselling may help explain possible causes and future pregnancy options. A fetal echocardiogram may be offered in a later pregnancy.

Living with it and what to expect

The first months may involve hospital care, surgery, medicines, and close monitoring of feeding and growth. Some babies need higher-calorie feeds or temporary feeding support.

After recovery, many children attend school, join family activities, and remain physically active. Safe activity depends on heart function, oxygen levels, rhythm, and the repair. The congenital cardiology team should provide individual advice.

Follow-up must continue throughout life, even when the person feels well. Narrowing, valve leakage, weak heart muscle, or abnormal rhythms can develop gradually without obvious symptoms. Adults should see a cardiologist trained in adult congenital heart conditions.

Some people need only monitoring for long periods; others need medicines, catheter procedures, or more surgery. Conduits placed in childhood do not grow with the body and may eventually require replacement.

Pregnancy increases demand on the heart. Anyone with repaired DORV who is considering pregnancy should have an assessment by an adult congenital heart and specialist pregnancy team before becoming pregnant.

Many people now live into adulthood. The outlook depends on the original anatomy, other health conditions, type of repair, heart function, and later complications.

When to get medical help

Call emergency services if a baby, child, or adult has:

Follow any emergency plan provided by the congenital heart team.

Seek prompt medical advice for increased breathing rate, feeding difficulty, sweating during feeds, reduced urine output, new swelling, worsening tiredness, poor weight gain, or a new or persistent fever.

Contact the congenital heart team soon about new palpitations, reduced exercise ability, dizziness, increasing breathlessness, or swelling. Keep all routine follow-up appointments even when there are no symptoms.

- Severe difficulty breathing, gasping, or pauses in breathing
- New, worsening, or severe blue or grey lips or tongue
- Collapse, fainting, or difficulty waking
- Sudden severe weakness or confusion
- Chest pain with fainting, severe breathlessness, or a very fast heartbeat

Common questions

Is double outlet right ventricle the same as a VSD?

No. A VSD is a hole between the lower heart chambers. In DORV, both main arteries connect mainly to the right ventricle. A VSD is usually present and allows blood to leave the left ventricle.

Can a baby survive double outlet right ventricle?

Many babies survive with surgery and specialist care. The outlook depends on the exact anatomy, other congenital conditions, heart function, and treatment results.

Will my child need more than one operation?

Some children have one main repair; others need staged operations. Later procedures may treat a narrowed pathway, aging conduit, valve problem, or another change.

Can someone with repaired DORV exercise?

Many people can be active, but safe intensity is individual. Competitive or high-intensity exercise may require assessment based on heart function, oxygen level, rhythm, and surgical history.

Is double outlet right ventricle genetic?

Usually, no single genetic cause is found. DORV can occur with a chromosome or gene difference, so genetics assessment may be useful when there are other congenital differences or a family history.

Can double outlet right ventricle be completely fixed?

Surgery can create effective circulation and improve survival, but lifelong monitoring remains necessary because repaired pathways, valves, heart muscle, and rhythm can change.

Sources

- Centers for Disease Control and Prevention. "Double Outlet Right Ventricle." Birth Defects
- American Heart Association. "Double Outlet Right Ventricle (DORV)." Congenital Heart Defects
- MedlinePlus Medical Encyclopedia, US National Library of Medicine. "Double Outlet Right Ventricle"
- Children's Hospital of Philadelphia. "Double Outlet Right Ventricle (DORV)." Cardiac Center
- Stout KK, Daniels CJ, Aboulhosn JA, et al. "2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease." Circulation . 2019;139:e698-e800
- Baumgartner H, De Backer J, Babu-Narayan SV, et al. "2020 ESC Guidelines for the Management of Adult Congenital Heart Disease." European Heart Journal . 2021;42:563-645