CONDITION

Double inlet left ventricle

Double inlet left ventricle is a heart condition present from birth in which one ventricle does most of the pumping.

WHAT IT IS
Congenital heart condition with one main pumping chamber

BODY SYSTEM
Heart and circulation

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

WHAT CAUSES IT
Abnormal heart development before birth, usually unexplained

HOW IT IS DIAGNOSED
Fetal or postnatal echocardiogram

MAIN TREATMENT
Staged heart surgery and lifelong specialist care

CAN IT BE CURED?
No - surgery reroutes circulation, not anatomy

IS IT AN EMERGENCY?
Sometimes - worsening blueness or severe breathing difficulty

LONG-TERM OUTLOOK
Lifelong complex heart condition; many survive into adulthood

Overview

Double inlet left ventricle is a complex heart condition that develops before birth. It is a single-ventricle heart condition , meaning one lower heart chamber does most of the pumping.

A typical heart has two upper chambers, called atria, and two lower chambers, called ventricles. The right ventricle pumps blood to the lungs, while the left ventricle pumps oxygen-rich blood to the body.

In double inlet left ventricle, both upper chambers connect mainly to the left ventricle. The right ventricle is usually small and may serve as an outlet chamber rather than a full pumping chamber. Oxygen-rich blood from the lungs and oxygen-poor blood from the body therefore mix inside the heart.

The effects depend on how easily blood reaches the lungs and body. Too little blood flow to the lungs can cause low oxygen levels and blue skin. Too much can lead to heart failure. Blood leaving the heart may also be obstructed.

Treatment usually involves several operations that redirect blood flow so the single working ventricle pumps mainly to the body. Surgery improves circulation but does not create a typical two-ventricle heart or remove the underlying condition.

Symptoms

Symptoms may begin within hours or days of birth, although their severity varies.

When too little blood reaches the lungs, signs can include:

When too much blood reaches the lungs, a baby may breathe quickly, sweat or tire during feeds, gain weight slowly, or develop swelling. The liver may become enlarged.

A heart murmur may be heard, but it does not indicate how serious the condition is. Some babies with severe heart disease have no obvious murmur.

After surgery, oxygen levels may remain lower than usual, particularly between treatment stages. Older children and adults may have reduced stamina, breathlessness, palpitations, swelling, or a bluish colour during exertion.

- Blue, grey, or dusky lips, tongue, or skin
- Fast or difficult breathing
- Low oxygen levels
- Unusual sleepiness
- Poor feeding

Causes and risk factors

Double inlet left ventricle results from altered heart development early in pregnancy. The structures that normally separate and connect the heart chambers do not form in the usual way.

In most cases, there is no identifiable cause. It is not caused by something a parent did or did not do during pregnancy.

Congenital heart conditions can sometimes be associated with a genetic or chromosome difference, but many affected people have no genetic explanation or family history.

A clinical genetics assessment may be offered when there are other physical findings, developmental concerns, or a family history of congenital heart disease. Genetic counselling can discuss testing and the chance of a heart condition occurring in a future pregnancy.

Diagnosis

Double inlet left ventricle may be found during pregnancy on a routine ultrasound. A fetal echocardiogram , a detailed ultrasound of the baby's heart, examines its chambers, valves, arteries, and blood flow.

After birth, symptoms or oxygen screening may lead to testing. An echocardiogram confirms the anatomy and shows how blood moves through the heart.

Other tests may include an electrocardiogram, chest X-ray, cardiac MRI, or CT scan. Cardiac catheterisation is sometimes needed to measure pressures and oxygen levels or obtain detailed images. In some cases, treatment can be performed during the same procedure.

Treatment and management

Treatment depends on the heart's anatomy and how blood reaches the lungs and body. Care is provided by a paediatric congenital heart team.

A newborn whose circulation depends on the ductus arteriosus , a fetal blood vessel, may receive medicine in hospital to keep it open temporarily. Breathing support, feeding support, and treatment for heart failure may also be required.

Some babies need an early procedure to balance blood flow. If too little blood reaches the lungs, a shunt or another procedure may provide a more reliable route. If too much reaches the lungs, a band may be placed around the pulmonary artery. Obstructed blood flow to the body may require a different operation.

Further surgery is generally performed in stages during infancy and early childhood. Common stages include:

After a Fontan procedure, blood returning from the body flows to the lungs without being pumped there by a ventricle. The single ventricle pumps oxygen-rich blood to the body.

These operations are palliative : they improve circulation but do not cure the condition. Surgical pathways vary. Heart transplantation may be considered if the single ventricle or Fontan circulation cannot be adequately supported.

Long-term care may include medicines, catheter procedures, rhythm treatment, further

surgery, nutritional care, and developmental support.

Good dental care helps reduce the risk of infection involving the heart. Antibiotics like amoxicillin or doxycycline before dental work are needed only in certain circumstances; the congenital heart team should confirm whether they are recommended.

- A bidirectional Glenn procedure, directing blood from the upper body to the lungs
- A Fontan procedure, directing blood from the lower body to the lungs

Complications

Lifelong monitoring is necessary because the single ventricle and surgically created circulation remain under unusual strain.

Possible complications include:

Some people need a pacemaker or treatment for a rapid heart rhythm.

Fontan circulation can also affect other organs. Raised pressure in the veins may contribute to liver scarring, kidney problems, abdominal fluid, or swelling. Uncommon but serious complications include protein-losing enteropathy, in which protein leaks through the bowel, and plastic bronchitis, in which firm material forms in the airways.

Exercise capacity is often below average. Some children also have difficulties with growth, learning, attention, or movement. Early support may help.

Follow-up may include blood tests, heart imaging, rhythm monitoring, exercise testing, and liver assessment so that changes can be found before major symptoms develop.

- Abnormal heart rhythms
- Heart valve leakage
- Reduced pumping strength or heart failure
- Blood clots
- Low oxygen levels
- Swelling or fluid accumulation

Prevention

Double inlet left ventricle usually cannot be prevented because it forms early in pregnancy and its cause is often unknown.

If you or your child has the condition and you are planning a pregnancy, genetic counselling may help determine whether testing is appropriate. A fetal echocardiogram may be offered in a future pregnancy.

For people living with the condition, preventive care includes attending congenital heart appointments, maintaining dental health, receiving recommended vaccines, avoiding smoking, and checking with the care team before starting medicines or supplements that could affect the heart or circulation.

Living with it and what to expect

Double inlet left ventricle is lifelong. Surgery changes blood flow, but the heart remains a single-ventricle heart. Specialist follow-up must continue even when the person feels well.

Many children attend school and participate in family and community life. Some need support with feeding, growth, learning, movement, or emotional wellbeing. Physical activity is often encouraged within limits set by the congenital heart team, although highly intense or competitive exercise may not be suitable.

Oxygen levels often improve after staged surgery, but stamina may remain reduced. Rhythm problems, valve leakage, ventricular weakness, or effects on the liver and other organs can develop over time, although their timing and severity vary.

Care should transfer from paediatric services to an adult congenital heart disease centre during adolescence or early adulthood. Routine adult cardiology services may not have the specialist experience required to manage Fontan circulation.

Pregnancy places substantial extra demands on the heart and can carry serious risks for someone with Fontan circulation. Pregnancy may be possible for some people, but planning should begin before conception with adult congenital heart and high-risk pregnancy specialists. Some may be advised that pregnancy is unsafe.

Many people now survive into adulthood. Long-term outlook depends on the exact anatomy, heart function, rhythm, surgical results, and Fontan-related complications. Some eventually need major additional treatment or transplant assessment.

When to get medical help

Call emergency services for:

For a baby awaiting or recovering from surgery, follow the heart centre's individual emergency

plan. Contact the specialist team promptly if feeding falls sharply, breathing becomes faster, the baby is difficult to wake, there are fewer wet nappies, or oxygen readings move outside the advised range.

Arrange prompt medical review for new or increasing swelling, abdominal enlargement, unexplained weight gain, ongoing diarrhoea, worsening tiredness, reduced exercise tolerance, frequent palpitations, or a persistent cough. These symptoms may indicate a circulation or Fontan-related problem.

Keep all routine congenital heart appointments, even when you or your child feels well. Rhythm, heart, or liver changes may not cause early symptoms.

- Severe difficulty breathing
- Collapse, unresponsiveness, or a seizure
- Blue or grey colouring that is sudden or much worse than usual
- Chest pain with fainting, severe breathlessness, or a racing heartbeat
- Sudden weakness, facial drooping, or trouble speaking

Common questions

Can double inlet left ventricle be cured?

No. Operations reroute blood flow but do not create a typical two-ventricle heart. Treatment can greatly improve circulation, and monitoring helps manage later problems.

Is double inlet left ventricle the same as hypoplastic left heart syndrome?

No. Both are single-ventricle conditions, but their anatomy differs. In double inlet left ventricle, the left ventricle is usually the main pumping chamber. In hypoplastic left heart syndrome, the left-sided pumping structures are severely underdeveloped.

Can a baby with double inlet left ventricle survive?

Many children survive staged surgery and reach adulthood. Outcomes depend on the anatomy, other health conditions, response to surgery, and later complications. The congenital heart team can explain an individual child's outlook.

Is double inlet left ventricle inherited?

It is not usually linked to one inherited change. Genetic factors may contribute in some families or when other birth differences are present. A genetics service can assess family history and discuss testing.

Can someone with this condition exercise?

Many people can and should remain active, but safe intensity varies. A congenital heart specialist can use symptoms and exercise testing to recommend suitable activities and limits.

Will someone with double inlet left ventricle need a heart transplant?

Not everyone does. Transplantation may be considered if the single ventricle weakens, Fontan circulation fails, or serious complications cannot be controlled. Assessment includes the heart and other organs, particularly the liver.

Sources

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