CONDITION

Ectopia cordis

Ectopia cordis is a birth condition in which the heart develops partly or completely outside the chest.

WHAT IT IS
Congenital condition with heart partly or fully outside chest

BODY SYSTEM
Heart and circulation, chest wall, and sometimes abdomen

MAIN SYMPTOMS
Exposed heart, breathing difficulty, blue skin, poor circulation

WHAT CAUSES IT
Early fetal chest-wall development disruption, usually unknown

HOW IT IS DIAGNOSED
Prenatal ultrasound, fetal echocardiography, or examination at birth

MAIN TREATMENT
Intensive newborn care and complex reconstructive surgery

CAN IT BE CURED?
Sometimes - surgery can successfully enclose heart

IS IT AN EMERGENCY?
Yes - exposed heart needs immediate newborn care

LONG-TERM OUTLOOK
High risk, shaped by location and associated defects

Overview

Ectopia cordis is a rare condition present from birth. The heart lies partly or completely outside the chest because the breastbone and nearby tissues did not close around it during early fetal development.

The heart may be covered by skin or a thin membrane, or it may be fully exposed. It can be positioned in the neck, chest, chest and abdomen, or abdomen. Many babies also have congenital heart defects or differences affecting the breastbone, diaphragm, abdominal wall, lungs, or abdominal organs.

Some babies have ectopia cordis as part of pentalogy of Cantrell , a group of midline birth differences involving the heart, breastbone, diaphragm, membrane around the heart, and upper abdominal wall. Incomplete forms do not include all five features.

An exposed heart can lose heat and fluid, become infected, or be injured. Abnormal heart structure and limited chest space may also interfere with breathing and circulation. When the condition is diagnosed before birth, specialist planning helps the team prepare for delivery and immediate treatment.

Symptoms

Ectopia cordis does not usually cause symptoms for the pregnant person and is often first found during prenatal ultrasound.

After birth, the main feature is a heart seen beating outside the chest or beneath a thin covering. Other signs may include:

Some symptoms result from an associated heart defect rather than the heart's position. Severity varies widely. A partly covered defect may be more manageable than a completely exposed heart with several major abnormalities.

- Fast, difficult, or weak breathing
- Blue or grey lips and skin from low blood oxygen
- Cool skin, low blood pressure, or other signs of poor circulation
- Feeding difficulty, tiredness, or poor weight gain
- An opening or swelling in the upper abdomen
- A short, split, or partly missing breastbone

Causes and risk factors

Ectopia cordis begins very early in pregnancy, when tissues that normally move towards the body's centre and form the chest wall do not join completely. The reason is usually unknown.

The condition is not caused by something a parent did or did not do during pregnancy. In most cases, no preventable exposure or behaviour is identified.

Chromosome or gene differences have been found in some affected pregnancies, especially when several organ systems are involved. However, most cases do not follow a clear inherited pattern and appear to happen sporadically.

The chance of recurrence is not well defined and may depend on whether testing identifies an underlying genetic cause. A clinical genetics team can review the baby's features, family history, and test results.

Diagnosis

Ectopia cordis may be detected by prenatal ultrasound, sometimes in the first trimester. Ultrasound can show the heart outside its expected position and an opening in the chest or abdominal wall.

A detailed fetal ultrasound checks the chest, abdomen, brain, limbs, and other organs. Fetal echocardiography , a specialised ultrasound of the heart, examines its chambers, valves, major blood vessels, rhythm, and blood flow. Fetal MRI may provide additional information about the lungs, diaphragm, chest, and abdominal organs.

Chromosome or broader genetic testing may be offered, particularly if other developmental differences are present. Testing can use a placental sample, amniotic fluid, or blood after birth. A normal result does not exclude every genetic cause.

After birth, examination confirms the heart's position. Echocardiography and other imaging assess internal heart structure, circulation, lung development, chest size, and associated abnormalities. These findings guide treatment and surgical planning.

Treatment and management

Care involves a specialist team that may include neonatal intensive care doctors, congenital heart and paediatric surgeons, cardiologists, anaesthetists, plastic surgeons, genetic specialists, and nurses.

When ectopia cordis is diagnosed before birth, delivery is usually planned at a centre with advanced newborn and surgical care. The delivery method depends on the pregnancy and the heart's size and position.

Immediately after birth, the team protects the heart, maintains temperature and fluids, supports breathing, and stabilises circulation. Exposed tissue may be covered with a sterile, non-stick dressing. Pressure, twisting, or attempts to push the heart into the chest must be avoided because they can damage or obstruct major blood vessels.

Some babies need mechanical ventilation, medicines to support circulation, or treatment for a congenital heart defect. Nutrition may initially be provided through a vein or feeding tube.

Surgery is tailored to the baby's anatomy. Its goals may include:

Placing the heart into a chest that is too small can compress the heart or lungs. Repair may therefore occur in stages, beginning with temporary coverage and followed by reconstruction when the baby is larger or more stable.

Some abnormalities are too extensive for successful repair. The team should explain the likely benefits and burdens of treatment. Comfort-focused care may be discussed when surgery cannot offer a reasonable chance of survival.

- Covering the heart with skin or other tissue
- Moving the heart into the chest when there is enough space
- Rebuilding the breastbone, chest wall, or abdominal wall
- Repairing defects inside the heart
- Closing openings in the diaphragm or membrane around the heart

Complications

Complications may result from the exposed heart, associated defects, limited chest space, or treatment. They include:

A reconstructed chest wall may grow differently and affect movement or breathing. Children who spend a long time in intensive care may need support with feeding, mobility, communication, hearing, or development.

- Injury, heat loss, fluid loss, or infection
- Low blood oxygen or inadequate circulation
- Breathing failure due to small lungs or chest-wall abnormalities
- Abnormal heart rhythms
- Compression of the heart or major blood vessels during reconstruction
- Bleeding, wound problems, or infection after surgery
- Feeding problems, slow growth, and the need for further operations

Prevention

There is no established way to prevent ectopia cordis. It develops very early in pregnancy, and its cause is usually unknown. Parents should not blame themselves for the diagnosis.

After an affected pregnancy or birth, genetic counselling can review test results and discuss early ultrasound or other testing in a future pregnancy. These measures support assessment and early detection; they do not prevent the condition.

Living with it and what to expect

The outlook depends on how much of the heart is outside the body, whether it has protective covering, the amount of chest space, and the severity of associated heart and organ abnormalities.

Ectopia cordis has a high risk of death, especially when the heart is completely exposed or several major abnormalities are present. Survival is possible when the heart can be protected, circulation is stable, and reconstruction is feasible. General survival figures may not accurately reflect an individual baby's anatomy.

Babies who survive the newborn period may remain in hospital for weeks or months and may need staged operations over several years. Follow-up usually monitors heart and lung function, chest-wall growth, nutrition, development, medicines, and surgical wounds.

After discharge, care may include heart medicines, feeding support, physical or occupational therapy, and temporary activity limits after surgery. Routine appointments remain important because abnormal rhythms, narrowing near a repair, or chest-wall restriction can develop gradually.

Social workers, psychologists, palliative care teams, and peer-support organisations can provide practical and emotional support. Palliative care can be given alongside active treatment and does not automatically mean other care is stopping.

When to get medical help

An exposed heart at birth is an emergency requiring immediate specialist newborn care. If ectopia cordis is unexpectedly present during a birth outside hospital, call emergency medical services at once. Do not push the heart into the chest or handle it unnecessarily.

A prenatal diagnosis does not usually place the pregnant person in immediate danger, but timely referral to fetal medicine and congenital heart specialists is essential for testing and delivery planning.

After hospital discharge, call emergency services for:

Contact the child's specialist team promptly for fever, increasing wound redness, drainage or swelling, worsening breathlessness, repeated vomiting, poor feeding, fewer wet nappies, unusual sleepiness, or a clear fall in activity. Follow the discharge plan about symptoms needing same-day assessment.

Continue routine follow-up for growth, development, heart and lung function, medicines, and future surgery. Do not wait for the next appointment if new concerns arise.

- Blue or grey lips, face, or skin
- Severe breathing difficulty, pauses in breathing, or collapse
- Unresponsiveness or extreme difficulty waking
- Heavy bleeding or sudden opening of a surgical wound
- Sudden severe paleness, cold skin, or other signs of worsening circulation

Common questions

Can a baby survive ectopia cordis?

Yes, but the condition is life-threatening. The outlook is generally better when the heart has some covering, there is enough chest space, and associated abnormalities can be treated. Imaging and the baby's condition after birth provide more useful guidance than general statistics.

Can the heart be put back inside the chest?

Sometimes. Surgeons assess chest size, lung position, and the course of major blood vessels first. Moving the heart into a chest that is too small can compress it, so gradual or staged reconstruction may be safer.

Is ectopia cordis the same as pentalogy of Cantrell?

No. Ectopia cordis can occur alone or with other abnormalities. Pentalogy of Cantrell is a broader group of five related midline defects, one of which may place the heart partly or completely outside the chest.

Will it happen again in another pregnancy?

Most cases have no clear inherited pattern, so an exact recurrence chance often cannot be given. Genetic counselling can interpret family history and any chromosome or gene test results.

Can ectopia cordis be seen before birth?

Yes. Prenatal ultrasound can often show the heart outside its expected position. Fetal echocardiography and sometimes fetal MRI help define the anatomy and plan care.

Does ectopia cordis hurt the baby?

Pain cannot be determined from appearance alone. Newborn teams provide pain relief and comfort care during procedures, ventilation, dressing changes, and surgical recovery. Pain and distress should be assessed regularly.

Sources

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