CONDITION

Tricuspid atresia

Tricuspid atresia is a birth defect in which the valve between the heart's right-sided chambers did not form.

WHAT IT IS
Birth defect with no tricuspid valve

BODY SYSTEM
Heart and circulation

MAIN SYMPTOMS
Blue-grey skin, fast breathing, feeding difficulty, poor growth

WHAT CAUSES IT
Heart development difference before birth, usually unknown

HOW IT IS DIAGNOSED
Fetal or postnatal echocardiogram

MAIN TREATMENT
Staged heart surgery and lifelong specialist care

CAN IT BE CURED?
No - surgery reroutes blood flow

IS IT AN EMERGENCY?
Sometimes - worsening blue colour or breathing difficulty

LONG-TERM OUTLOOK
Many reach adulthood but need lifelong specialist monitoring

Overview

Tricuspid atresia is a heart condition present from birth. The tricuspid valve, which should connect the heart's upper-right chamber to its lower-right chamber, has not formed. A solid layer of tissue is present instead.

This means blood cannot follow its usual route through the right side of the heart and into the lungs. The lower-right chamber, called the right ventricle, is usually smaller than normal because little or no blood enters it.

A baby can survive only if blood can take other routes through the heart. These usually include an opening between the upper chambers and another opening or blood vessel that allows blood to reach the lungs. Blood low in oxygen then mixes with oxygen-rich blood, which can cause a blue or grey colour known as cyanosis .

Tricuspid atresia is often described as a single-ventricle heart condition because one main pumping chamber must do most of the work. Surgery can redirect blood flow and improve oxygen delivery, but it does not create a normal tricuspid valve or a second working ventricle. Lifelong specialist heart care remains important.

Symptoms

Symptoms often begin at birth or during the first days or weeks of life. Their timing and severity are affected by how much blood reaches the lungs and how freely blood moves between the heart's chambers.

Possible symptoms in a baby include:

Changes in skin colour may be easier to see on the lips, tongue, gums and around the eyes in people with darker skin.

If too much blood flows to the lungs, a baby may develop signs of heart failure. In babies, heart failure means the heart cannot keep up with the body's needs; it does not mean the heart has stopped. Signs can include rapid breathing, sweating with feeds, slow growth and swelling.

Older children and adults may have reduced exercise capacity, tiredness, shortness of breath, a bluish colour, irregular heartbeats or swelling in the abdomen, legs or ankles. Clubbing, where the fingertips become broader and rounder, can develop after long periods of low blood oxygen.

- Blue or grey lips, tongue, skin or nail beds
- Fast or difficult breathing
- Tiring, sweating or becoming breathless during feeds
- Taking a long time to feed or drinking less than expected
- Poor weight gain
- Unusual sleepiness or low energy

Causes and risk factors

Tricuspid atresia develops early in pregnancy while the baby's heart is forming. In most cases, it is not clear why the valve fails to develop.

Changes in genes may contribute in some people, and the condition occasionally occurs with a genetic syndrome or other birth differences. However, it is usually not caused by a single

inherited gene. Having one child with a congenital heart condition may change the chance of it happening in another pregnancy, but the individual chance requires specialist assessment.

Nothing a parent did or did not do can usually be identified as the cause. Tricuspid atresia may occur alongside other heart differences, including holes between heart chambers, narrowing near the lungs or changes in the positions of the main arteries.

Diagnosis

Tricuspid atresia may be suspected during a routine pregnancy ultrasound. A fetal echocardiogram , which is a detailed ultrasound of the baby's heart, can examine the valves, chambers and blood flow. Some cases are not found until after birth.

After birth, low oxygen levels on newborn pulse-oximetry screening, a blue-grey colour or an unusual heart sound may lead to further testing. A pulse oximeter is a painless sensor placed on the skin to estimate oxygen in the blood.

An echocardiogram usually confirms the diagnosis. This ultrasound shows that the tricuspid valve is absent and identifies the openings through which blood is moving. It also checks the heart's pumping function and looks for other structural differences.

Other tests may include:

During catheterisation, a thin tube is passed through a blood vessel to the heart. It may be used before surgery or later to investigate problems and sometimes to treat them.

- An electrocardiogram to record the heart's electrical activity
- A chest X-ray to assess the heart and lungs
- Cardiac MRI or CT for more detailed images
- Cardiac catheterisation to measure pressures and examine blood flow

Treatment and management

A newborn with tricuspid atresia needs prompt assessment by a specialist congenital heart team. Early care focuses on keeping enough blood flowing to the lungs and maintaining safe oxygen levels.

Some babies need medicine to keep a fetal blood vessel called the ductus arteriosus open until a procedure can be performed. If the opening between the upper heart chambers is too small, a catheter procedure called balloon atrial septostomy may enlarge it.

Most children have staged operations. The exact sequence is based on the heart's anatomy and blood flow.

An early operation may create a route from an artery to the lungs when too little blood reaches them. If blood flow to the lungs is excessive, a different procedure may be used to limit it.

Later operations usually connect the large veins directly to the lung arteries. The final stage is commonly called the Fontan procedure . After this operation, blood returning from the body flows toward the lungs without being pumped there by a ventricle. The main working ventricle pumps oxygen-rich blood around the body.

These operations are called palliative surgery . Here, palliative means that surgery improves circulation without restoring a two-ventricle heart. It does not mean that care is only for the end of life.

Long-term management may include medicines to support heart function, control heart rhythm or reduce the chance of blood clots. These are chosen individually. Some people need further catheter procedures, surgery, a pacemaker or, in advanced cases, assessment for heart transplantation.

Regular care at a congenital heart centre is important even when you feel well. Follow-up may include oxygen measurements, blood tests, heart imaging, rhythm monitoring, exercise testing and checks of the liver and other organs.

Complications

Tricuspid atresia and the circulation created by surgery place lasting demands on the heart and other organs. Complications do not occur in everyone, and they may appear years after surgery.

Possible complications include:

Some people also have learning, movement or attention difficulties related to complex congenital heart disease and early treatment. Developmental assessment and school support can help when these concerns arise.

Infective endocarditis is an infection of the heart's inner lining. Antibiotics like amoxicillin or doxycycline before dental procedures are recommended only for certain heart conditions and circumstances. A congenital heart specialist or dentist can explain whether this applies to you.

- Irregular heart rhythms
- Reduced heart-pumping function or heart failure

- Blood clots, including clots that may travel to the lungs or brain
- Narrowing or blockage in surgically created pathways
- Fluid retention and swelling
- Liver scarring caused by long-term pressure in the veins
- Protein-losing enteropathy, in which protein leaks through the bowel
- Plastic bronchitis, in which firm material forms in the airways
- Reduced exercise capacity

Prevention

Tricuspid atresia usually cannot be prevented because the reason it develops is not known.

If you have tricuspid atresia, have had a child with it or have a family history of congenital heart conditions, genetic counselling may help explain possible recurrence risks. During a future pregnancy, fetal echocardiography may be offered to examine the developing heart. This is a screening step, not a guarantee that a heart condition will or will not occur.

Living with it and what to expect

Most children need several procedures in infancy and early childhood, followed by regular monitoring throughout life. Hospital visits may become less frequent during stable periods, but follow-up should not stop. Changes in heart rhythm, circulation, liver health or exercise tolerance can begin before obvious symptoms appear.

Many people with treated tricuspid atresia attend school, work and take part in family and social life. Physical stamina is often lower than in people with a typical heart. Suitable activity is usually encouraged, but safe limits should be discussed with a congenital heart team because they are not the same for everyone.

Good dental care is especially important because mouth infections can enter the bloodstream. Regular dental visits, brushing and treatment of dental problems help reduce this risk.

Pregnancy places substantial extra strain on a Fontan circulation. Anyone considering pregnancy should have pre-pregnancy assessment through adult congenital heart and high-risk pregnancy services. Some people can carry a pregnancy, while others face a high risk of complications.

The long-term course is not identical for everyone. Some remain stable for many years, while

others need treatment for rhythm problems, heart failure, liver changes or failing Fontan circulation. Continuing care with an adult congenital heart specialist gives the best chance of detecting these changes early.

When to get medical help

Call emergency services for:

If a child has an established care plan, follow its emergency instructions while seeking help.

Contact the congenital heart team promptly for a new fast or irregular heartbeat, increasing shortness of breath, reduced exercise ability, rapid weight gain, persistent cough, abdominal swelling or swelling of the legs and ankles. Fever with unexplained tiredness or worsening heart symptoms also needs prompt advice.

Routine follow-up is appropriate when symptoms are stable and no new concerns have appeared. Keep scheduled appointments even if you or your child feels well.

- Severe trouble breathing
- A sudden or marked increase in blue or grey colour
- Fainting, collapse or difficulty waking
- New weakness on one side, facial drooping or trouble speaking
- Severe chest pain, especially with sweating or breathlessness
- A baby who is blue, struggling to breathe or unable to feed

Common questions

Is tricuspid atresia the same as a missing heart chamber?

No. The tricuspid valve is missing, and the right ventricle is usually underdeveloped rather than completely absent. The other chambers are present, although there are usually additional openings that allow blood to mix and reach the lungs.

Can tricuspid atresia be cured with surgery?

Surgery can greatly improve blood flow, oxygen levels and survival, but it does not rebuild a normal valve or create two working ventricles. The Fontan circulation requires lifelong monitoring, so tricuspid atresia is not considered cured.

How many surgeries are needed?

Most children have more than one operation, usually in stages. The number and timing are influenced by the heart's structure, lung blood flow, previous procedures and later complications. Catheter procedures may also be needed.

Can someone with tricuspid atresia exercise?

Many people can and should remain physically active within limits set for their circulation. Walking, swimming or other moderate activity may be suitable, while intense competitive or heavy resistance exercise may not be safe for some people. An individual exercise assessment is useful.

Can an adult with tricuspid atresia have children?

Biological parenthood may be possible. Pregnancy can carry major risks for someone with a Fontan circulation, including heart rhythm problems, heart failure, blood clots and pregnancy complications. Assessment before pregnancy is essential. Medicines may also need review because some are unsafe during pregnancy.

What is the life expectancy with tricuspid atresia?

There is no single life expectancy that applies to everyone. Many people now survive into adulthood after staged surgery, but long-term complications remain possible. Outlook is influenced by heart function, rhythm, oxygen level, Fontan circulation, liver health and access to specialist follow-up.

Sources

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- American Heart Association: Evaluation and Management of the Child and Adult With Fontan Circulation, 2019 scientific statement
- American Heart Association and American College of Cardiology: 2018 Guideline for the Management of Adults With Congenital Heart Disease