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## Heart disorders (acquired) - children

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### Summary

- An acquired heart defect develops in children after a disease.
  - There are four types of acquired heart defect.
  - All children with heart defects have special needs when taking medication.
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Some children acquire a heart problem after an illness in childhood. This is called an acquired heart defect. This is different to a congenital (at birth) heart defect, which occurs when a child is born with a heart problem.

The four main types of acquired heart disorder in children are:

- Kawasaki disease – an illness that occurs mainly in young children and may leave the heart muscle or coronary arteries damaged.
- Myocarditis – the heart muscle becomes inflamed and may be damaged after a viral infection.
- Cardiomyopathy – a disease of the heart muscle, caused by a genetic disorder or after an infection. It leads to poor heart function.
- Rheumatic heart disease – caused by rheumatic fever, this disease leads to heart muscle and valve damage.

### Kawasaki disease

Kawasaki disease usually, but not always, affects children aged five years and under. It may damage the heart muscle or coronary arteries. It is named after the Japanese paediatrician who identified the disease. The cause is unknown but the disease is probably caused by an abnormal reaction to a common germ. Kawasaki disease is not contagious, although it can occur in clusters.

### Symptoms of Kawasaki disease

The main symptom of Kawasaki disease is persistent fever (over 38.5°C) for five days or longer. There is usually no obvious explanation for the fever and it generally does not respond to paracetamol. Other symptoms, usually caused by inflammation of small blood vessels known as vasculitis, may include:

- A rash, sometimes in the groin region
- Red, swollen and cracked lips
- Red eyes
- Bright red, swollen tongue
- Swollen hands and feet
- Red rash on palms of hands and soles of feet
- Swollen lymph nodes.

### Diagnosis of Kawasaki disease

There is no test to diagnose Kawasaki disease. Diagnosis is made by excluding other possible causes of symptoms. Diagnosis may require blood tests and an echocardiogram to examine the heart for any changes in the coronary arteries.

Most children who have Kawasaki disease and receive proper treatment will make a full recovery. A few children will develop heart problems, including damage to the coronary arteries. If no treatment is given, about 25 per cent of patients experience inflammation of the coronary arteries, which supply blood to the heart muscle. This can cause irregularities with these blood vessels, which may disturb the flow of blood.

### **Treatment for Kawasaki disease**

The treatment for Kawasaki disease is intravenous gammaglobulin (immunoglobulin), made from donated blood transfusions. Large doses of intravenous gammaglobulin will usually stop the fever and other symptoms of Kawasaki disease. Treatment should be administered within 10 days of the onset of fever to minimise heart problems.

Children may also be prescribed aspirin for some weeks following the onset of Kawasaki disease, to prevent problems with coronary arteries. However, aspirin should only be given to children on the advice of a doctor, paediatrician or cardiologist. Aspirin is not usually recommended for children because of the risk of Reye's syndrome, a rare but potentially fatal disease.

### **Myocarditis**

Myocarditis is an inflammation of the heart muscle. Infections can damage the heart's cells. The normal immune system response is to attack the organism. However, in some children this immune system attack is too aggressive and destroys heart muscle cells as well as the 'foreign' organism.

The heart muscle can thicken and swell as a result of this immune response. The damaged heart muscle cells may heal, or may form scar tissue. If a large part of the heart is affected, its ability to pump blood may be impaired.

Autoimmune diseases, medications and chemicals can trigger myocarditis. However, in most children myocarditis is triggered by an infection, usually viral. Possible causes of infection include:

- Influenza (flu)
- Glandular fever
- Rheumatic fever
- Rubella
- HIV
- Diphtheria.

### **Symptoms of myocarditis**

There is no specific test for myocarditis, and there may be no obvious symptoms. Symptoms, when they do occur, may not be easy to detect. Symptoms may include:

- Poor circulation, showing as cold hands and feet
- Skin discolouration in blue or grey tones
- Decreased urine production, due to impaired kidney function
- Chest pain and palpitations
- Fever
- Swelling in the face, feet or legs.

### **Diagnosis of myocarditis**

Tests used to diagnose myocarditis may include:

- X-rays, including chest x-rays. This may show whether the heart is enlarged or if there is fluid in the lungs
- Electrocardiogram
- Echocardiogram or heart ultrasound
- Blood tests to assess kidney and liver function
- Heart biopsy – this requires a catheter to be inserted through a leg blood vessel to obtain a tiny piece of heart muscle.

Many children will recover completely from myocarditis, but some may develop significant heart failure. In rare cases, patients can develop blood clots that lead to stroke or heart attack, or may develop serious heart

arrhythmia, which can be fatal.

### **Treatment for myocarditis**

There is no cure for myocarditis. Treatment focuses on the underlying cause, as well as supporting the heart to function and maintain adequate circulation. Treatment can include:

- Medication to control blood pressure and body fluids (diuretics)
- Intravenous immunoglobulin or purified antibodies, to reduce inflammation
- Bed rest and avoiding strenuous physical activity.

Immunisations against the viral diseases that can trigger myocarditis, including rubella and influenza, are an important way to prevent this and other acquired heart disease in children.

### **Cardiomyopathy**

Cardiomyopathy is a disease of the heart muscle. There are three main types:

- Dilated cardiomyopathy – an enlargement of one or more of the heart's chambers
- Hypertrophic cardiomyopathy – a thickening of the heart's muscle
- Restrictive cardiomyopathy – the heart muscle becomes more rigid.

In most cases of cardiomyopathy in children, the cause is unknown. However, possible factors include:

- Heart valve problems
- Viral infections that trigger myocarditis
- A family history of cardiomyopathy
- Genetic disorders including Noonan syndrome.

### **Symptoms of cardiomyopathy**

Symptoms of cardiomyopathy vary. Some people, including children, have no symptoms in the earlier stages. Symptoms, where they occur, may include:

- Swelling of the hands and feet
- Swelling of the abdomen with fluid
- Breathlessness
- Fatigue
- Irregular heart rhythm
- Dizziness, light-headedness and fainting during physical activity.

### **Diagnosis of cardiomyopathy**

Tests to diagnose cardiomyopathy may include:

- Chest x-ray
- Electrocardiogram
- Echocardiogram
- Blood tests
- Cardiac catheterisation.

### **Treatment for cardiomyopathy**

Treatment will depend on the type of cardiomyopathy diagnosed. Treatment may include:

- Medications – angiotensin-converting enzyme (ACE) inhibitors, diuretics and beta blockers to improve the heart's pumping capability
- Devices – a pacemaker to coordinate the contractions between the left and right ventricle, or an implantable cardioverter defibrillator (ICD) to monitor the heart rhythm and deliver electrical shocks to control abnormal, rapid heartbeats

- Surgery and transplant – a heart transplant, which may be an option for patients with advanced symptoms or who have not been successfully treated by other measures.

## **Rheumatic heart disease**

Rheumatic heart disease is caused by acute rheumatic fever. This disease can cause long-term damage to the heart muscle or heart valves, especially repeated and untreated episodes. Early diagnosis and treatment of acute rheumatic fever can prevent rheumatic heart disease.

Acute rheumatic fever is a delayed complication of streptococcal disease, a throat infection with group A streptococcus bacteria. Only a small number of people infected with the bacteria will develop acute rheumatic fever, but the risk increases if the infection is not treated, usually with penicillin.

Acute rheumatic fever is rare in Australia, except among Aboriginal and Torres Strait Islander children and young people where the rate of infection is high. The incidence of acute rheumatic fever in Aboriginal and Torres Strait Islander children aged between five and 14 years varies between 250 and 350 per 100,000 of population.

### **Symptoms of acute rheumatic fever**

Symptoms of acute rheumatic fever can include:

- Fever
- Joint pain and swelling
- Carditis (inflammation of the heart).

### **Treatment for acute rheumatic fever**

Acute rheumatic fever is treated with medication, usually antibiotics including penicillin. Children with the fever are usually hospitalised.

Where the disease has damaged heart valves (rheumatic heart disease), surgery may be needed to repair or replace damaged valves.

### **Heart problems and medication**

All children with heart defects should be given antibiotics when they have teeth removed or when they have other surgery which involves the mouth, nose, intestinal organs or genito-urinary systems. Bacteria may enter the bloodstream during these procedures and can cause serious infection (known as infective endocarditis) in the abnormal part of the heart.

All medications should be checked with your pharmacist, doctor or cardiologist. The usual immunisations should be given at the normal times after advice from your doctor.

### **Where to get help**

- Your family doctor
- NURSE-ON-CALL Tel. 1300 60 60 24 – for expert health information and advice (24 hours, 7 days)
- A paediatric cardiologist (your doctor can refer you)
- HeartKids Victoria Inc. Tel. (03) 9513 9030

### **Things to remember**

- An acquired heart defect develops in children after a disease.
- There are four types of acquired heart defect.
- All children with heart defects have special needs when taking medication.

**This page has been produced in consultation with and approved by:**

HeartKids Victoria

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