outline

REPAIRING A BABY'S BROKEN HEART

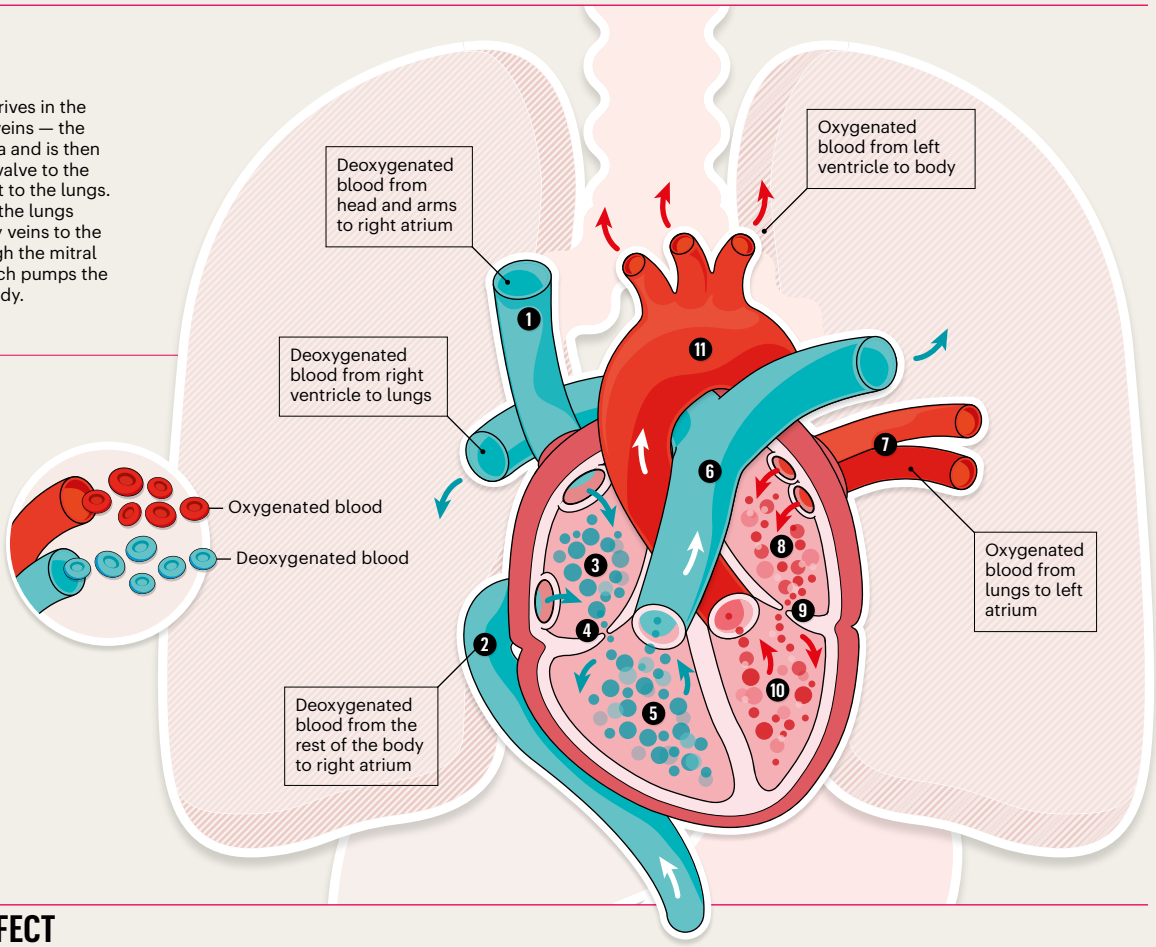
Some babies are born with a rare disorder in which only one of the lower chambers of their heart works properly. These single-ventricle defects (SVDs) can be managed through a complex series of operations. *By* **Benjamin Plackett**; *infographic by* **Alisdair Macdonald.**

HEALTHY HEART CIRCULATION

Deoxygenated blood (blue) arrives in the heart's right atrium from two veins — the superior and inferior vena cava and is then passed through the tricuspid valve to the right ventricle, which pumps it to the lungs. Oxygenated blood (red) from the lungs travels through the pulmonary veins to the heart's left atrium, then through the mitral valve to the left ventricle, which pumps the blood out to the rest of the body.

Anatomy

1. Superior vena cava (SVC)
2. Inferior vena cava (IVC)
3. Right atrium
4. Tricuspid valve
5. Right ventricle
6. Pulmonary artery
7. Pulmonary veins
8. Left atrium
9. Mitral valve
10. Left ventricle
11. Aorta



THREE KINDS OF DEFECT

The anatomy of each SVD differs, but they all result in a mix of oxygenated and deoxygenated blood being pumped to the body and lungs. This means that vital organs don't get enough oxygen.

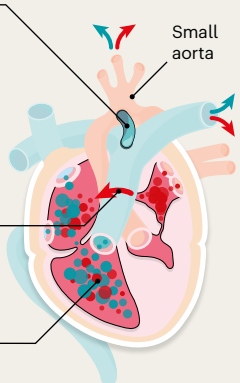
Survival rate for this condition[1]

Hypoplastic left heart syndrome (HLHS)
The left ventricle and aorta are underdeveloped.

Ductus arteriosus: this blood vessel connects the pulmonary artery and aorta. It usually closes after birth but needs to be kept open to allow some mixed blood to reach the body.

Atrial septal defect: a hole between the atria allows oxygenated blood to cross into the right atrium.

Oxygenated and deoxygenated blood mix.

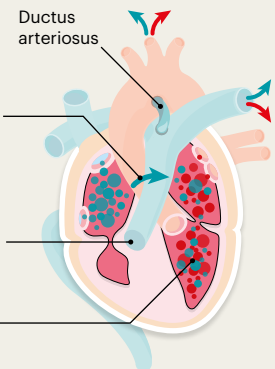


Pulmonary atresia
The right ventricle is underdeveloped and not connected to the pulmonary artery.

Atrial septal defect: a hole between the atria allows deoxygenated blood to cross into the left atrium.

Blood cannot pass from the right ventricle into the pulmonary artery and on to the lungs.

Oxygenated and deoxygenated blood mix.

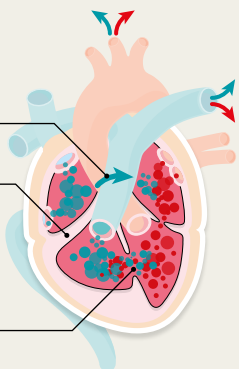


Tricuspid atresia
The right atrium isn't connected to the right ventricle and there is a hole between the ventricles.

Atrial septal defect: a hole between the atria allows deoxygenated blood to cross into left atrium.

Missing tricuspid valve.

Ventricular septal defect: a hole between the ventricles means mixed blood goes to the lungs and body.



Watch an animation at nature.com/collections/heart-defects-outline

A SOLUTION IN STAGES

SVDs are treated with a sequence of three operations[2]. Together, these surgeries ensure that the deoxygenated blood returning from the body bypasses the heart and goes straight to the lungs. The heart is then left to pump only oxygenated blood — getting rid of the problem of mixed blood.

Norwood procedure

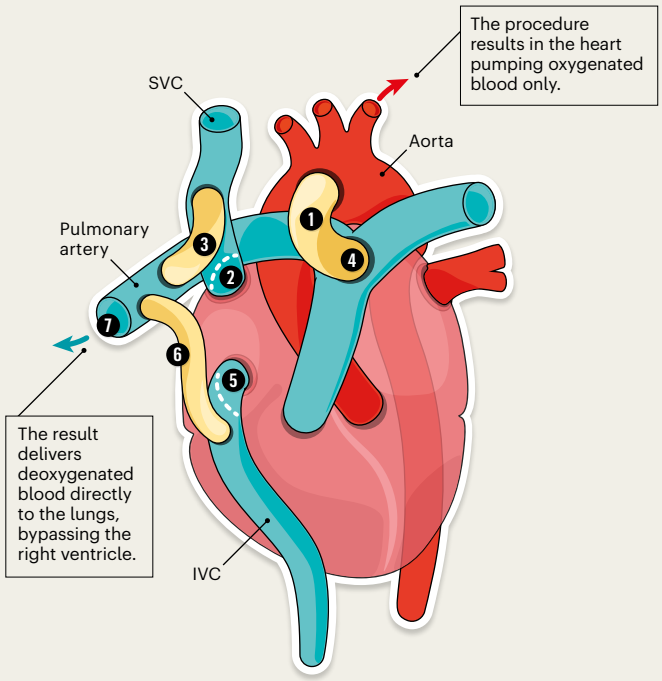
1. A Blalock–Taussig (BT) shunt tube replaces the ductus arteriosus. This allows oxygenated and deoxygenated blood to mix and reach the body.

Glenn procedure

2. The SVC is disconnected from the heart.
3. The SVC is connected directly to the pulmonary artery.
4. The BT shunt is removed.

Fontan completion

5. The IVC is disconnected from the heart.
6. The IVC is routed to the pulmonary artery.
7. Deoxygenated blood bypasses the heart entirely.

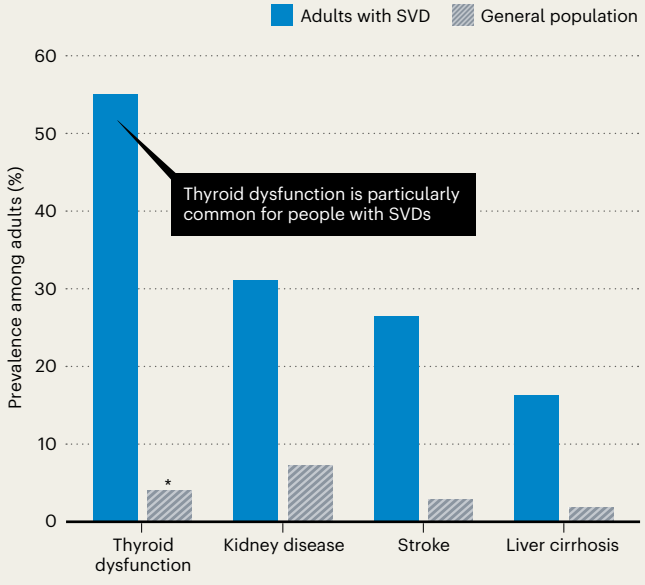


COMORBIDITIES ARE COMMON

Complications

Children born with SVDs can live into their 30s and 40s, but they have a significantly higher chance of experiencing other diseases as compared with healthier adults[3–7].

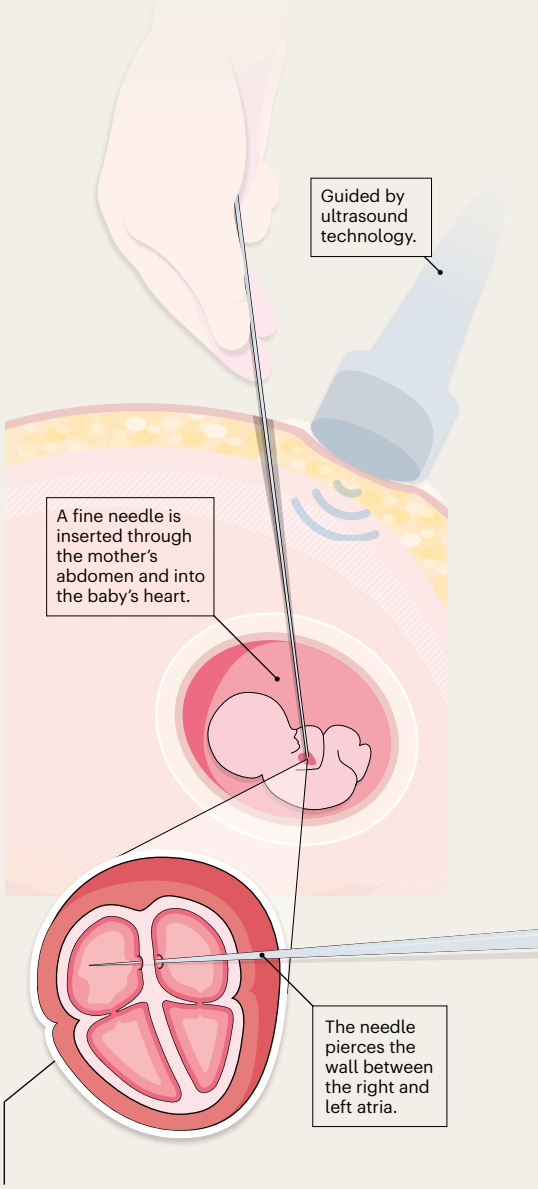
9 in every 100,000 people (roughly) have a type of single-ventricle defect[8].



*Data from ref. 4; all other data from studies of German populations refs 3 & 5–7.

IN UTERO SURGERY

Surgeons have begun to pioneer procedures to start treatment from within the womb.



Fetal balloon atrial septostomy

This helps the very few babies that have an SVD but no atrial septal defect. Surgeons insert a needle through the mother's abdomen and into the baby's heart. This punctures a hole in the atrial wall to allow oxygenated and deoxygenated blood to mix so the baby can survive long enough for corrective surgery.

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