

Supporting young people and adults
born with a heart condition

The
Somerville
Foundation

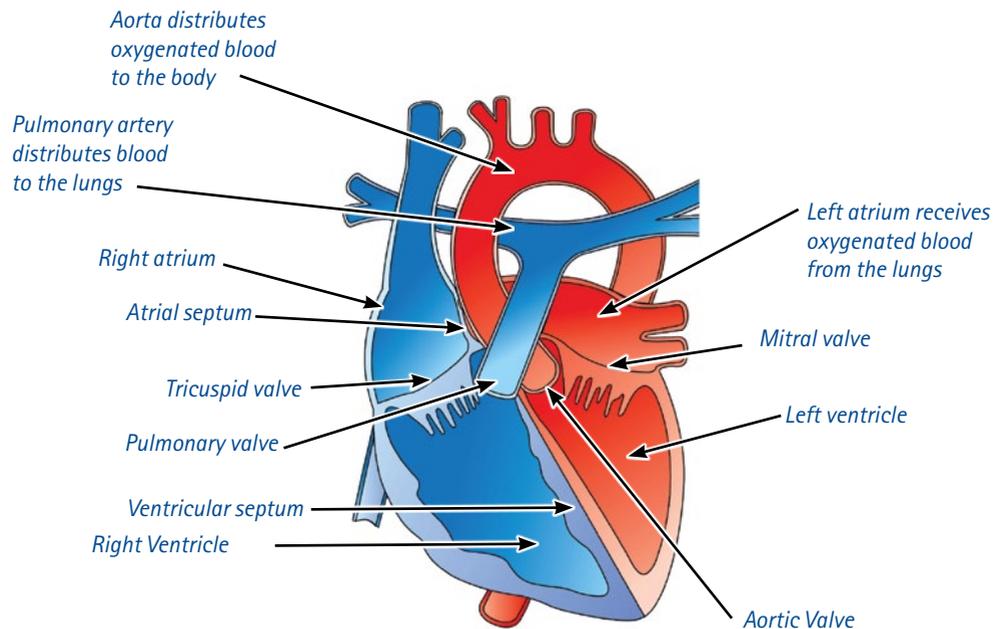


Tetralogy of Fallot (Repaired)

> Your heart, your life, your choices

The leaflet provides information to help you understand the heart condition with which you were born and to make choices about your treatment. Your medical team will work in partnership with you and help you make informed decisions. Remember though it is your heart, your life and ultimately your choices.

> The Normal Heart



> What Is Tetralogy Of Fallot?

Tetralogy of Fallot is a combination of four heart irregularities that are present from birth. The Greek word 'Tetra' means four and Fallot was the name of the French physician who first described the condition. Around 10% of people who are born with a heart condition have Tetralogy of Fallot.

Blue, deoxygenated blood returns to the right side of your heart from your body in the normal way. The blood flow is then disrupted by the following:

1. There is a narrowing in the outflow tract of the right ventricle that takes blood from the heart to the lungs. This is called pulmonary stenosis. This narrowing can range from mild to severe and may involve the pulmonary artery and its branches and could also restrict the pulmonary valve.
2. The muscle of the right ventricle becomes thicker than normal. This due to your right ventricle working harder (because of the pulmonary stenosis) to push deoxygenated blood through the narrow pulmonary valve and up to your lungs.

> Tetralogy Of Fallot (Unrepaired)

AO – Aorta

RA – Right Atrium

RV – Right Ventricle

LA – Left Atrium

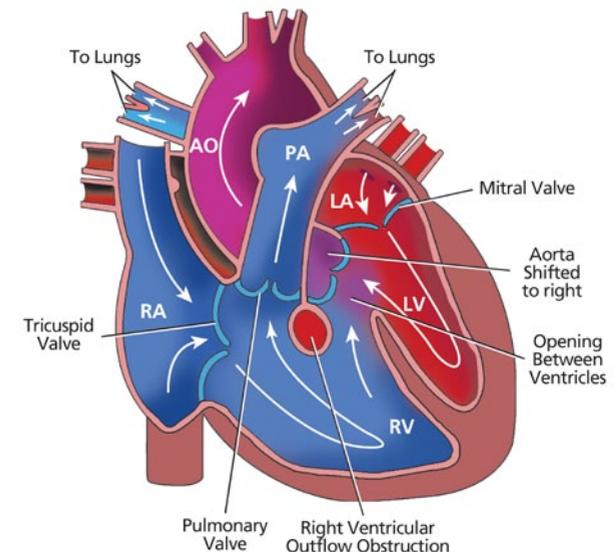
LV – Left Ventricle

PA – pulmonary Artery

Colour – Blue (de-oxygenated blood)

Pink (Oxygenated blood)

Purple (mixing of pink and blue blood)



3. There is a hole between the left and right ventricles (LV & RV) of your heart. This is called a ventricular septal defect (VSD). Some of the deoxygenated blood will pass across the VSD and mix with the pink oxygenated blood in the left ventricle (purple blood).
4. The aorta that takes red oxygenated blood around your body straddles the VSD (hole). This causes a mixture of oxygenated and deoxygenated blood to enter the aorta and flow around the body. This is known as an overriding aorta.

Depending on the severity of the narrowing on the right side, you may have appeared blue as a baby.

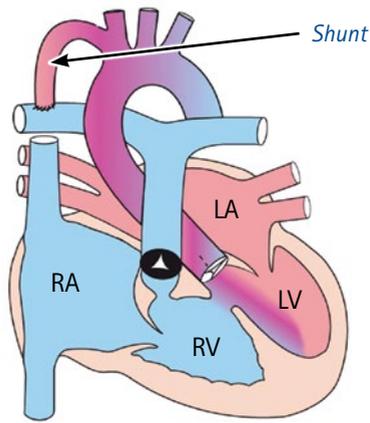
The two main issues are:

- The VSD (hole) between the left and right ventricles,
- The narrowing in the outflow tract or pulmonary valve (i.e. pulmonary stenosis).

> What operations did I have as a child?

You may have had a 'shunt' operation to increase the flow of blood to your lungs and have a scar on the side of your chest. A shunt is a palliative operation which means that it helped to relieve symptoms you had, rather than correcting the irregularity.

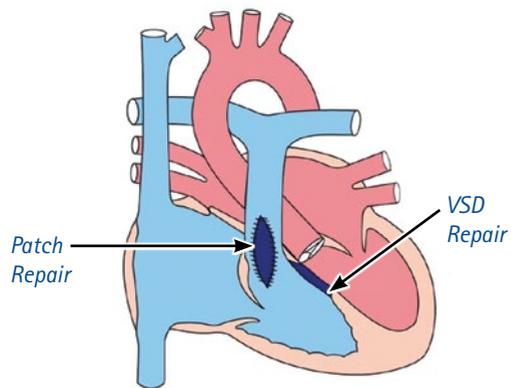
> Shunt Operation



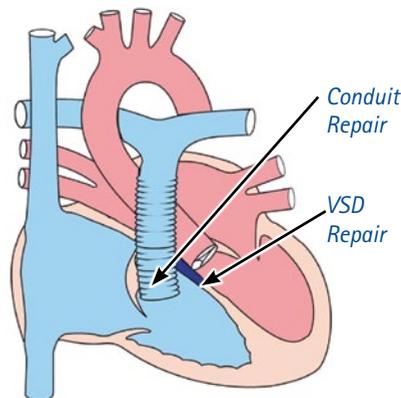
You may also have had a corrective procedure. The corrective operation to repair Tetralogy of Fallot has two main parts:

- Close the VSD (hole) between the left and right ventricles so that the blood from the left ventricle is directed to the aorta.
- Widen the narrowing of the pulmonary valve between the right ventricle and the pulmonary artery so that blood can flow more freely to the lungs. This was probably repaired in one of the following ways
 - a. Widening the narrow area by removing some thickened muscle that may have developed below the pulmonary valve
 - b. Widening the narrow area (i.e. the outflow tract or the pulmonary valve) with a Transannular patch (see diagram below)
 - c. Inserting a conduit, which is a new tube that contains a pulmonary valve to replace the narrowed vessel (see diagram below)
 - d. Replacing the pulmonary valve
 - e. Stretching the pulmonary valve.

> Repair Using a Transannular Patch



> Repair Using a Conduit



> Reasons for long term follow-up

It is important that you are followed up for life by a Cardiologist who specialises in Congenital Heart Disease as you may develop issues that could need medical intervention. These include:

Pulmonary valve incompetence:

- a. If you had a widening procedure your pulmonary valve may not work as efficiently as you get older, allowing deoxygenated blood to flow back through the valve. This is sometimes referred to as a 'leaky valve'.
- b. If you had a conduit inserted it can become narrowed or hardened with calcium over time, reducing the flow of blood to the lungs.

Further narrowing of the pulmonary arteries

- Problems may develop in the smaller blood vessels in the lungs called the distal pulmonary arteries. These may become narrowed in adulthood.

An irregular heartbeat

- You may develop palpitations or abnormal heart rhythms.
- Your right ventricle may become stretched over time as it has to work harder to pump deoxygenated blood to the lungs.
- The aorta may also show signs of stretching. This is not very common.

Your Cardiologist will discuss various options with you, including the possibility of further surgery, drug treatment or procedures to alleviate your symptoms. Depending on the choice you make regarding your treatment you may be referred to other specialists who have expertise in Congenital Heart matters.

> What tests will I need?

As well as an echocardiogram (ECHO) and electrocardiogram (ECG) your Cardiologist may also suggest one or more of the following tests that are used to assess the condition of your heart:

- Right and left heart catheter
- Cardiac Magnetic Resonance Imaging Scan
- 24hr ECG
- Cardio-pulmonary exercise test
- Specialised electrophysiology studies to examine the electrical pathways of your heart

> What treatment options could be available to me?

Your Specialist GUCH Consultant Cardiologist will discuss various options with you depending on your individual needs, for example:

You may require an operation to

- a. Replace the pulmonary valve
- b. Replace the conduit on the right side of the heart
- c. Repair or replace the aortic valve and blood vessel on the left side of the heart

Some treatments may be possible via a cardiac catheter procedure

- a. It may be possible to replace your pulmonary valve through keyhole surgery, but this is not suitable for everybody.
- b. It may be possible to expand the narrowed area within an artificial covering by inserting a small metal cage, called a stent, to keep the area open.
- c. It may also be possible to expand smaller lung arteries and keep them open by inserting a small stent

You may require special electrophysiology treatments for troublesome heart rhythms which are not controlled by medication.

You may require a pacemaker to speed up your heart rate.

> What symptoms should I tell my Cardiologist about?

It is important to inform your health care team such as your cardiologist, nurse or GP of any new symptoms as they occur.

- Decrease in exercise tolerance
- Increase in breathlessness
- Dizziness
- Fainting

It is advisable that you ask your specialist for an individualised management plan, developed by you and your GUCH medical team, to provide details of who and when to contact if you have any concerns or changes of symptoms or circumstances.

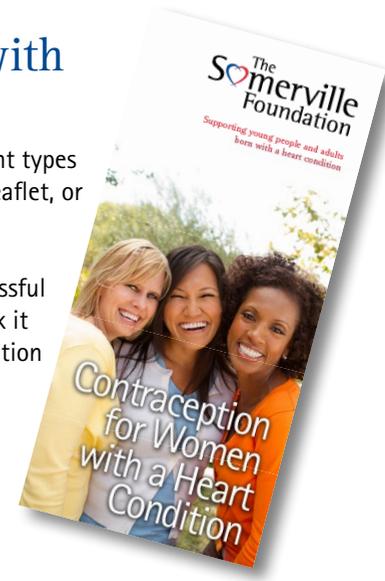
> Exercise with repaired Tetralogy of Fallot

There are few restrictions on the types of exercise and sports you will be able to enjoy and your specialist cardiologist will be able to help you make an informed decision regarding the level of exercise that is safe for you to undertake. Depending on your preference, it may be that you will need to have additional tests before and during your elected form of exercise which will be explained to you by your medical team.

> Contraception and Pregnancy with repaired Tetralogy of Fallot

The Somerville Foundation has produced a leaflet on the different types of contraception available. Please contact us for a copy of the leaflet, or visit www.thesf.org.uk and view the information online.

Many women with repaired Tetralogy of Fallot have had a successful pregnancy and a spontaneous normal delivery. To reduce any risk it is advisable to plan your pregnancy, taking into account information given to you by your congenital cardiac team. You should have the opportunity to enjoy a shared care arrangement during your pregnancy and delivery from both your Consultant Cardiologist and Consultant Obstetrician.



> Lifestyle choice

There are few restrictions on the types of jobs, travelling and life enhancing experiences that you may have.

> Life Assurance / Insurance

At the moment there are no long term survival tables for congenital cardiac conditions. Insurance companies rely on these types of tables to offer individual policies e.g. critical illness, life insurance and assurance relating to a mortgage. The Somerville Foundation is striving towards making insurance available for you. You may be able to obtain cover through your employer, or an insurance broker may be able to help, although this could result in a higher premium than normal.

Travel insurance is available, although sometimes at a higher premium than normal depending on your individual health issues. Download our Travel Insurance leaflet from www.thesf.org.uk or call The Somerville Foundation office on 01473 252007 for a copy.

The Somerville Foundation would like to thank the North West Adult Congenital Cardiac Service team for allowing us to reproduce information in this leaflet.



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