

Tetralogy of Fallot

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Learning objectives

- What is Tetralogy of Fallot (TOF)
- How TOF is treated
- Some of the issues patients with TOF may encounter

What is Tetralogy of Fallot?

- A type of congenital heart disease
- One of the most common cyanotic types of heart disease
- Consists of 4 defects that occur together

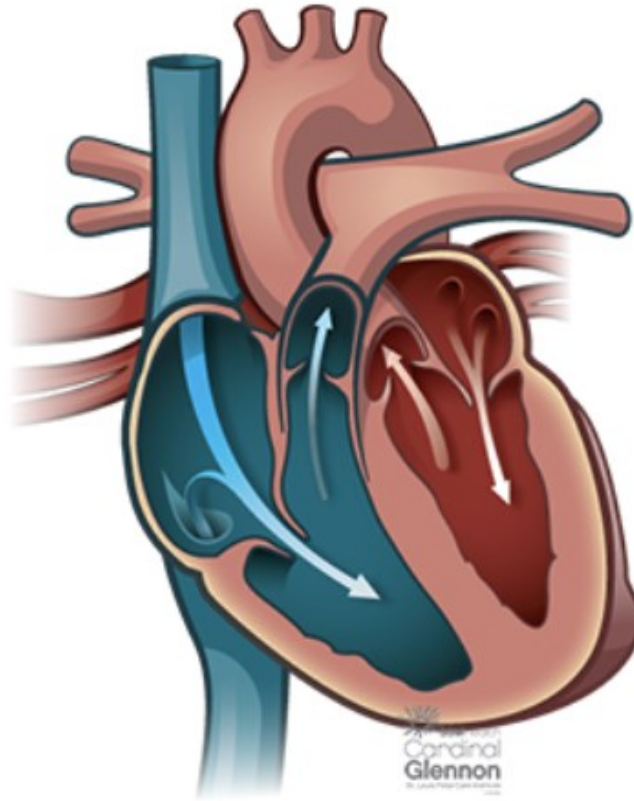
Tetralogy = denotes 4 parts.

Fallot = French physician

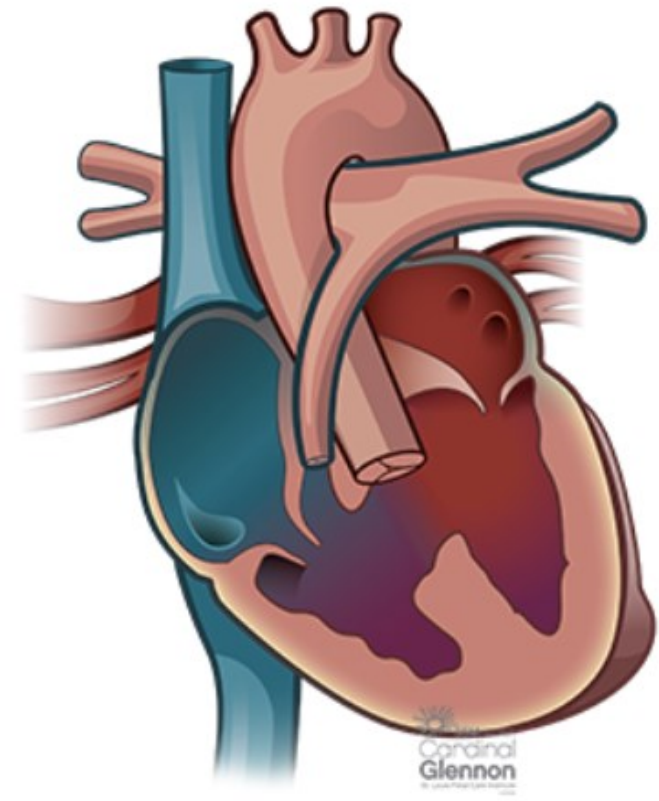


Tetralogy of Fallot

- Ventricular septal defect
- Overriding aorta
- Pulmonary stenosis
- Right ventricular hypertrophy



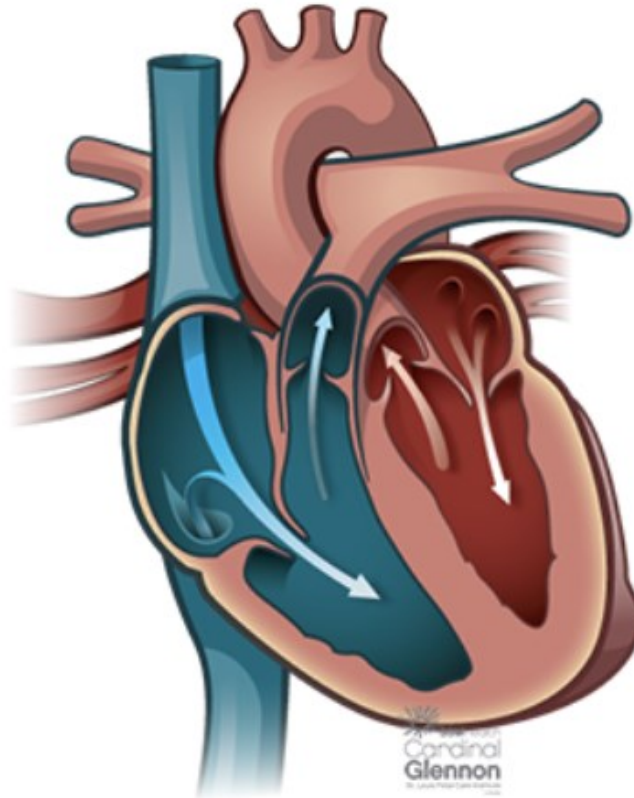
Healthy Heart



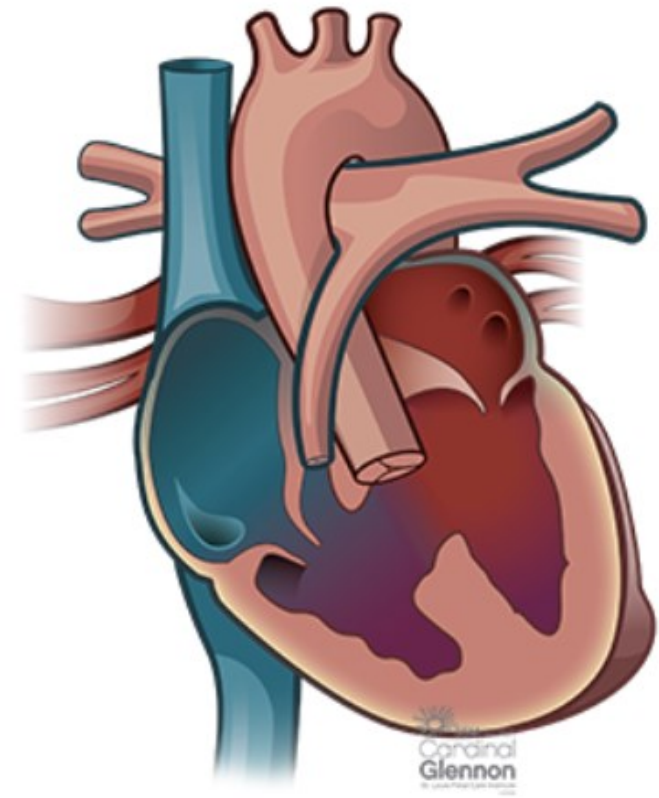
Tetralogy of
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Tetralogy of Fallot management

- Complete surgical repair as baby when big enough
- May need temporary measures to improve blood flow to lungs whilst waiting



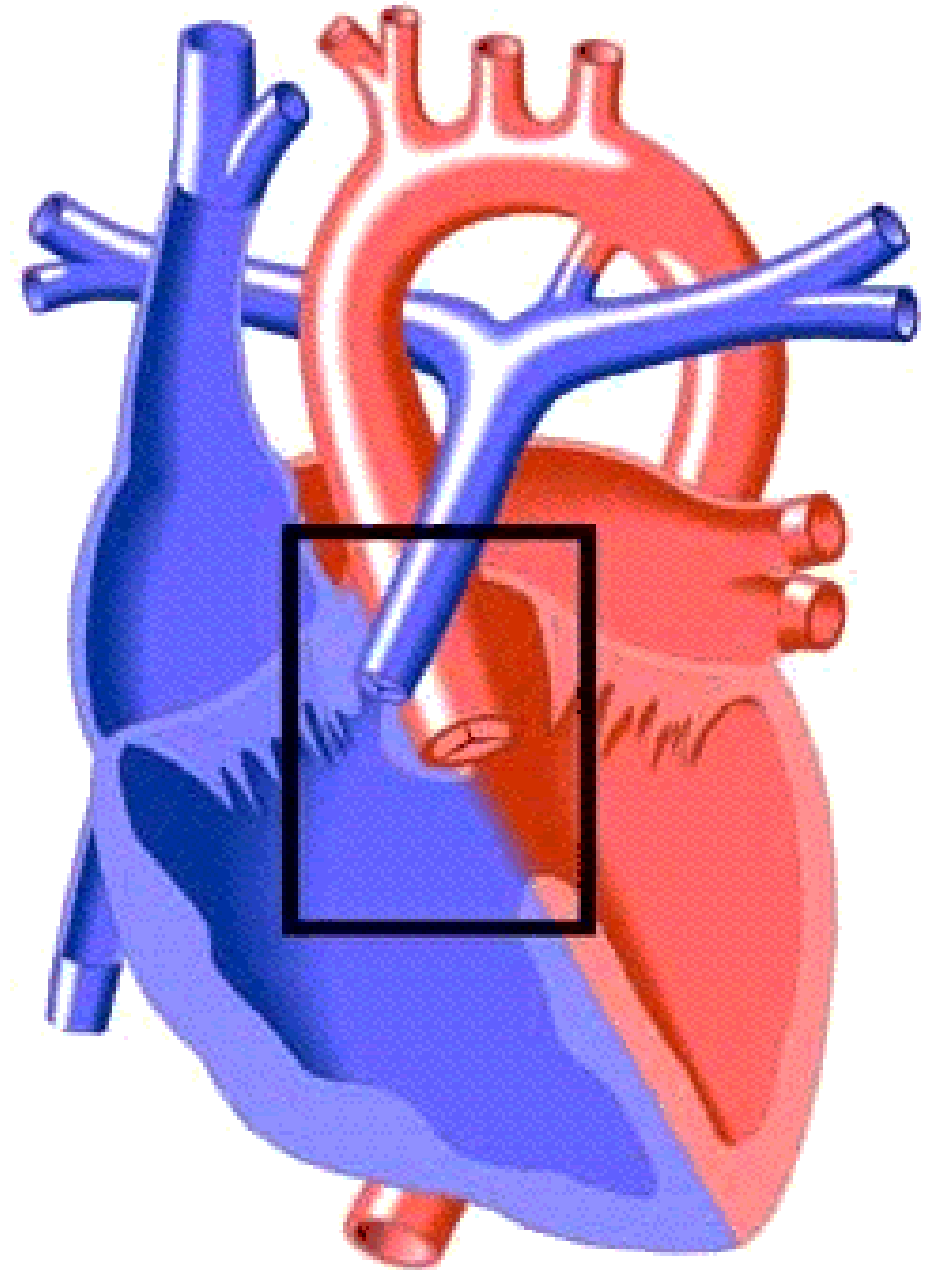
Healthy Heart



Tetralogy of
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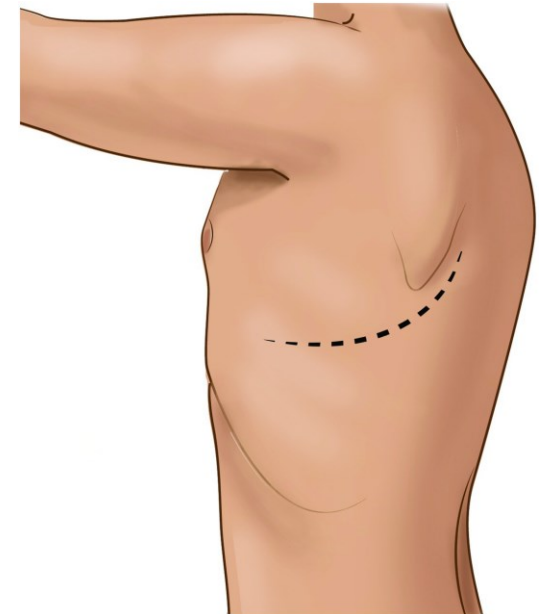
Tetralogy of Fallot repair

- Close the VSD
- Restore adequate blood flow to the lungs



An adult patient with repaired TOF

- Midline sternotomy scar
 - Should have normal saturations
 - Should have normal blood pressure/heart rate
 - Will probably have a murmur
 - Will probably have a RBBB on ECG
-
- Some patients may have lateral thoracotomy scar
(If there is a lateral thoracotomy scar,
check BP is equal on both sides)



<https://www.cambridge.org/core/books/abs/atlas-of-surgical-techniques-in-trauma/chest/454686463C0D62CCBFC074D28E69A3D1>

An adult patient with unrepaired TOF

- May be cyanotic
- May have clubbing
- May have heart failure
- May have heart rhythm disturbances



'Tet spell'

- Babies/young children with unrepaired TOF may have sudden episodes of profound cyanosis and hypoxia.
- Usually triggered by an event such as feeding or emotional upset.
- Thought to occur as a result of imbalance between pulmonary and systemic vascular resistance that leads to decreased pulmonary blood flow and increased right to left shunting.
- Placing knees to chest may help
- May need oxygen and medical therapy if severe

Prognosis

- Mortality rate in untreated patients reaches 50% by age 6 years.
- Now most people with repaired TOF enjoy long term survival with a good quality of life.
- Still do need long term follow up.



TOF complications

- Pulmonary valve regurgitation or stenosis
 - Most patients with TOF will need further intervention on the pulmonary valve
 - May be done either surgically or percutaneously, depending on anatomy.

2020 ESC Guidelines for the management of adult congenital heart disease

The Task Force for the management of adult congenital heart disease of the European Society of Cardiology (ESC)

Recommendations for intervention after repair of tetralogy of Fallot

Recommendations	Class ^a	Level ^b
PVRep is recommended in symptomatic patients with severe PR ^c and/or at least moderate RVOTO. ^d	I	C
In patients with no native outflow tract, ^e catheter intervention (TPVI) should be preferred if anatomically feasible.	I	C
PVRep should be considered in asymptomatic patients with severe PR and/or RVOTO when one of the following criteria is present. <ul style="list-style-type: none"> • Decrease in objective exercise capacity. • Progressive RV dilation to RVESVi ≥ 80 mL/m², and/or RVEDVi ≥ 160 mL/m² ^f, and/or progression of TR to at least moderate. • Progressive RV systolic dysfunction. • RVOTO with RVSP >80 mmHg. 	IIa	C

VSD closure should be considered in patients with residual VSD and significant LV volume overload or if the patient is undergoing pulmonary valve surgery.

IIa

C

In patients with sustained VT who are undergoing surgical PVRep or transcatheter valve insertion, pre-operative catheter mapping and transection of VT-related anatomical isthmuses before or during the intervention should be considered.

IIa

C

Electrophysiologic evaluation, including programmed electrical stimulation, should be considered for risk stratification for SCD in patients with additional risk factors (LV/RV dysfunction; non-sustained, symptomatic VT; QRS duration ≥ 180 ms, extensive RV scarring on CMR).

IIa

C

ICD implantation should be considered in selected TOF patients with multiple risk factors for SCD, including LV dysfunction, non-sustained, symptomatic VT, QRS duration ≥ 180 ms, extensive RV scarring on CMR, or inducible VT at programmed electrical stimulation.

IIa

C

Catheter ablation or concomitant surgical ablation for symptomatic monomorphic sustained VT may be considered in those with a preserved biventricular function as an alternative to ICD therapy, provided that the procedure is performed in highly experienced centres and that established ablation endpoints have been reached (e.g. non-inducibility, conduction block across ablation lines).

IIb

C

TOF complications

- Infective endocarditis
 - Usually affecting the pulmonary valve
 - All patients should be advised of ways to reduce their risk of endocarditis (regular dental reviews, avoiding tattoos and piercings, early treatment of infection)



TOF complications

- Heart rhythm disturbance
 - May get atrial or ventricular arrhythmias
 - This can be due to scar tissue as a result of their operation
 - May need a pacemaker or defibrillator

TOF complications

- Heart failure

Summary

- Tetralogy of Fallot is a cyanotic congenital heart disease
- Usually repaired in childhood
- Most patients will need further intervention or surgery on their pulmonary valve
- Should have normal observations
- May have serious complications

Thank you for listening

Any questions?