Final FRCA Teaching Tetralogy of Fallot

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ST6 Anaesthetics

Introduction

- Quick quiz - knowledge check
- Anatomy
- Physiology
- Clinical implication in children

Quiz - Slido

- **www.slido.com**
- **#194546**

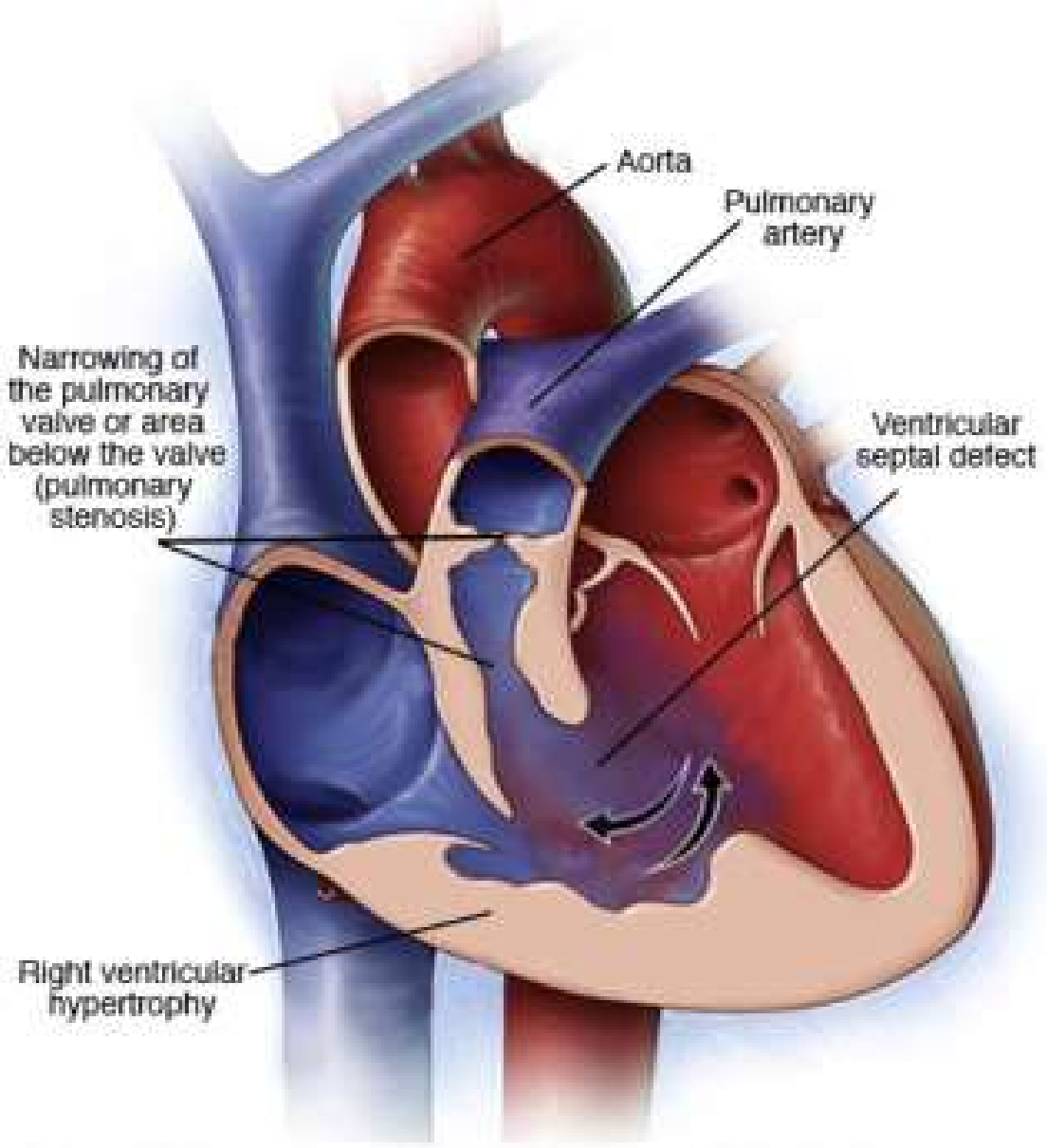
Tetralogy of Fallot

- Common cyanotic congenital heart disease
  - 1 in 3500 live births
  - Around 10% of congenital cardiac disease

- Names after French physician Etienne-Louis Fallot who reported the first case series in 1888.

Anatomy

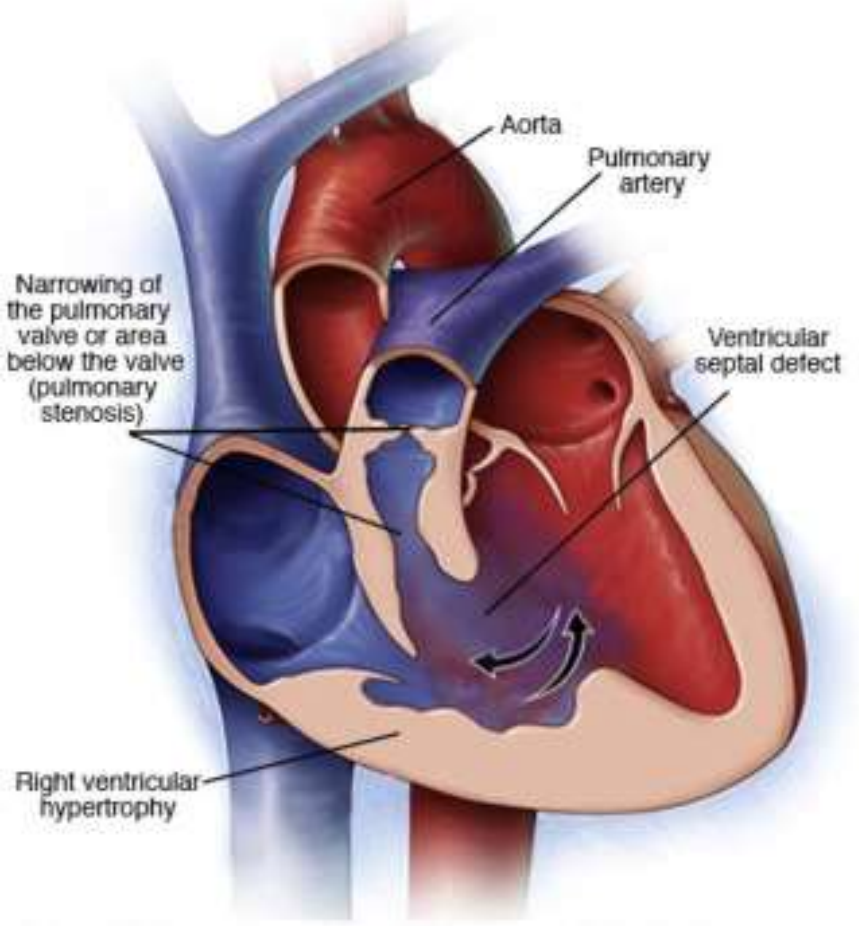
- VSD
- RVOTO
- Overriding aorta
- RVH



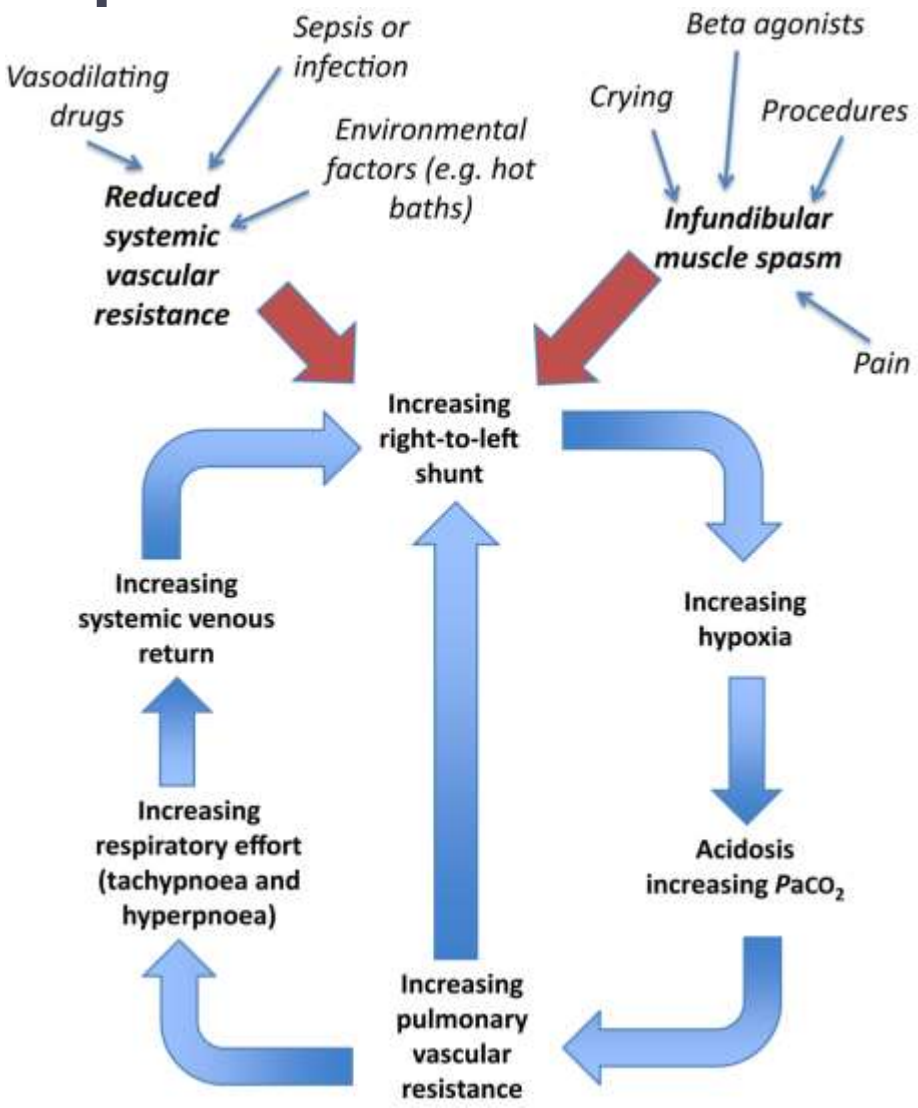
TOF variations

Pathophysiology

- Mixing of blood via the VSD
- Normally right to left shunt = cyanosis
- Shunt determined by pressure gradient between RV and LV
- Pulmonary blood flow determined by degree of RVOTO



Cyanotic spells



Clinical features

Majority are antenatal diagnosis on scan
Post-natally may be detected due to

- Cyanosis; worsens with age
- Hypoxia with minimal response to O2 therapy
- Heart murmur (pansystolic and ejection systolic)
- Spelling episodes
- Arrhythmias
- Associated genetic conditions (~30% cases)
  - Downs
  - DiGeorge
  - Alagille
  - VACTERL

Investigations

Investigation	Key findings
CXR	Boot shaped heart – RVH Reduced pulmonary vascular markings (RVOTO) Look for other causes of hypoxia Acyanotic – CXR may be like that of VSD with pulmonary oedema and normal pulmonary vascular markings
Blood Gas	Low PaO2 (not increased with O2) Signs of decompensation – lactate and bicarb
ECG	R axis deviation Tall R wave in V1; RVH Tall P waves (P Pulmonale); R atrial enlargement ?Arrhythmias
Echo	Confirm diagnosis and assess - Severity of RVOTO - Level of RVOTO - Intra-cardiac anatomy - Any associated congenital defects - Coronary artery anatomy
Cardiac catheterisation	Rarely needed unless in complex cases to assess coronaries, assess for collaterals or obtain pressure measurements

Pre repair care

- If severe RVOTO; maintain PDA to ensure adequate pulmonary blood flow
  - Prostin infusion
- Dynamic RVOTO; B-blockade may help

- Majority of ToF don't require active management and go home until large enough for corrective surgery

Spelling; acute deterioration

- Triggers
  - Sympathetic stimulation, including pain and anxiety (e.g. during venepuncture)
  - Exercise
  - Breath holding or Valsalva manoeuvre
  - Crying, feeding, and defaecation
  - Vasodilatation and decrease in SVR (e.g. hot baths)
  - Hypoxia
  - Hypercarbia
  - Acidosis
  - Induction of anaesthesia
  - Sympathomimetic drugs

Clinical features

- Restless
- Agitated
- Breathless
- Older children – squat position = increases SVR so reduces R-L shunt
- Cyanosis
- Tachycardia

Acute management

- Remove any precipitant if known
- Try to calm the child and provide comfort
- Knee-to-chest flexion position
- High-flow oxygen (may only improve saturations once the shunt fraction is reduced, but it helps to lower the PVR)
- Titrated sedation and analgesia (morphine, fentanyl, and ketamine)
- Fluid bolus to improve RV filling
- Increase the SVR to reduce the right-to-left shunt with α-agonists (phenylephrine and noradrenaline) or vasopressin

Acute management continued

- Avoid primarily beta-agonist sympathomimetics (e.g. dopamine and adrenaline) as these substances can worsen the infundibular spasms. However, adrenaline should be used in profound collapse if ventricular function is poor and coronary artery perfusion at risk (e.g. profound hypotension)
- Intubation, artificial ventilation, and use of neuromuscular blocking agents if no improvement or profound hypoxia
- Reduce $PaCO_2$ to decrease PVR.
- Consider esmolol or other β-blockers for persistent infundibular spasm and to slow down the HR to improve the diastolic filling time (expert use only)
- If all fails, urgent surgical intervention for either palliative or complete repair

Non-cardiac surgery; anaesthesia aims

- Normally done at regional paediatric cardiac centre
- Key principles
  - Avoid spell triggers
  - Avoid excessive SVR reductions
  - Avoid excessive PVR increases
  - Avoid excessive sympathetic stimulation

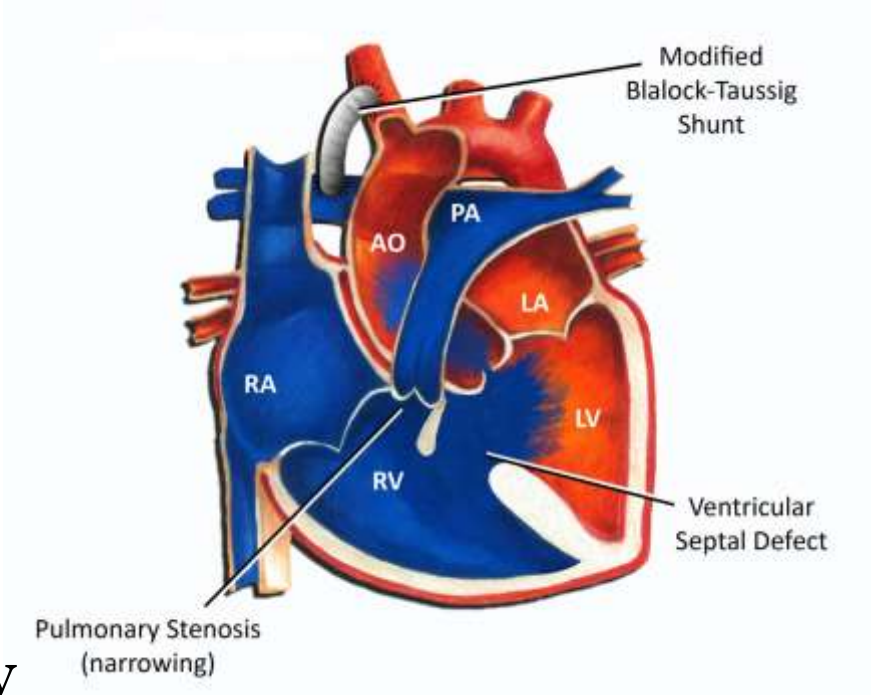
ToF procedures

Cardiac cath interventions

- Improve pulmonary blood flow whilst waiting for child to grow
- Pulmonary valvotomy or RVOT stenting
- PDA stenting

Temporising/Palliative surgery

- Systemic to pulmonary shunt created
- Modified Blalock-Taussig (mBT)
- Subclavian artery and ipsilateral pulmonary artery



Corrective surgery

- Performed around 6 months old
- Open cardiac surgery with bypass
  - Patch closure of the VSD
  - Aorta back to LV
  - Resection of RVOT muscle bundles
  - Reduction of the RVOT valvular stenosis
  - Repair of any associated findings eg ASD, PDA

Post repair management

- PICU
- General cardiac surgery complications
  - Bleeding
  - Systemic inflammatory response
- Specific complications
  - Arrhythmias (junctional ectopic tachycardia)
  - Residual lesions (VSD, RVOTO, Pul regurg)
  - Restrictive RV physiology
  - Pleural effusions (high RV end-diastolic pressure)

Long term

- Lifelong follow up
  - Can be poor compliance and patients may be unaware of their diagnosis
- Despite repair to form a biventricular system
  - Variable physisology
    - Pulmonary valve regurgitation
    - RV dysfunction
    - Residual lesions (tiny VSD, mild RVOTO)
- Pregnancy
  - Discussion with regional MDT advised!

Prognosis

- If uncorrected life expectancy is between 1st -4th decade of life
  - Affects quality of life, growth, development and educational achievement
  - Death often due to cardiac failure, infections, thromboembolic disease
- Corrected – good outcomes especially if no associated syndrome
  - Increased risk of sudden death; arrhythmias
  - May have ICD or ablation performed
  - Pulmonary valve incompetence may occur

Questions

References

- Tetralogy of Fallot. R. Wilson, O. Ross and M.J. Griksaitis. BJA Education, 19(11): 362e369 (2019)
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