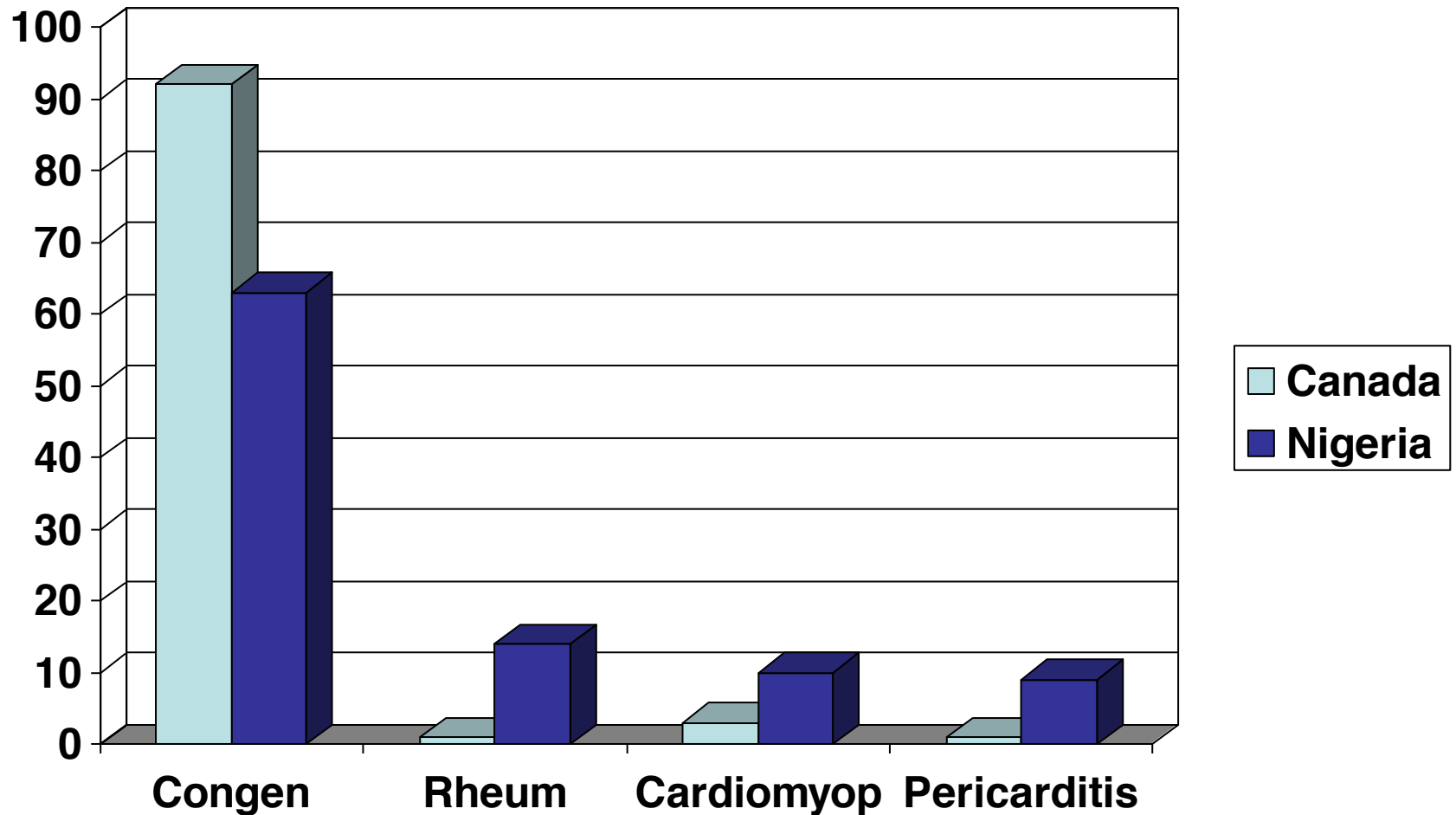


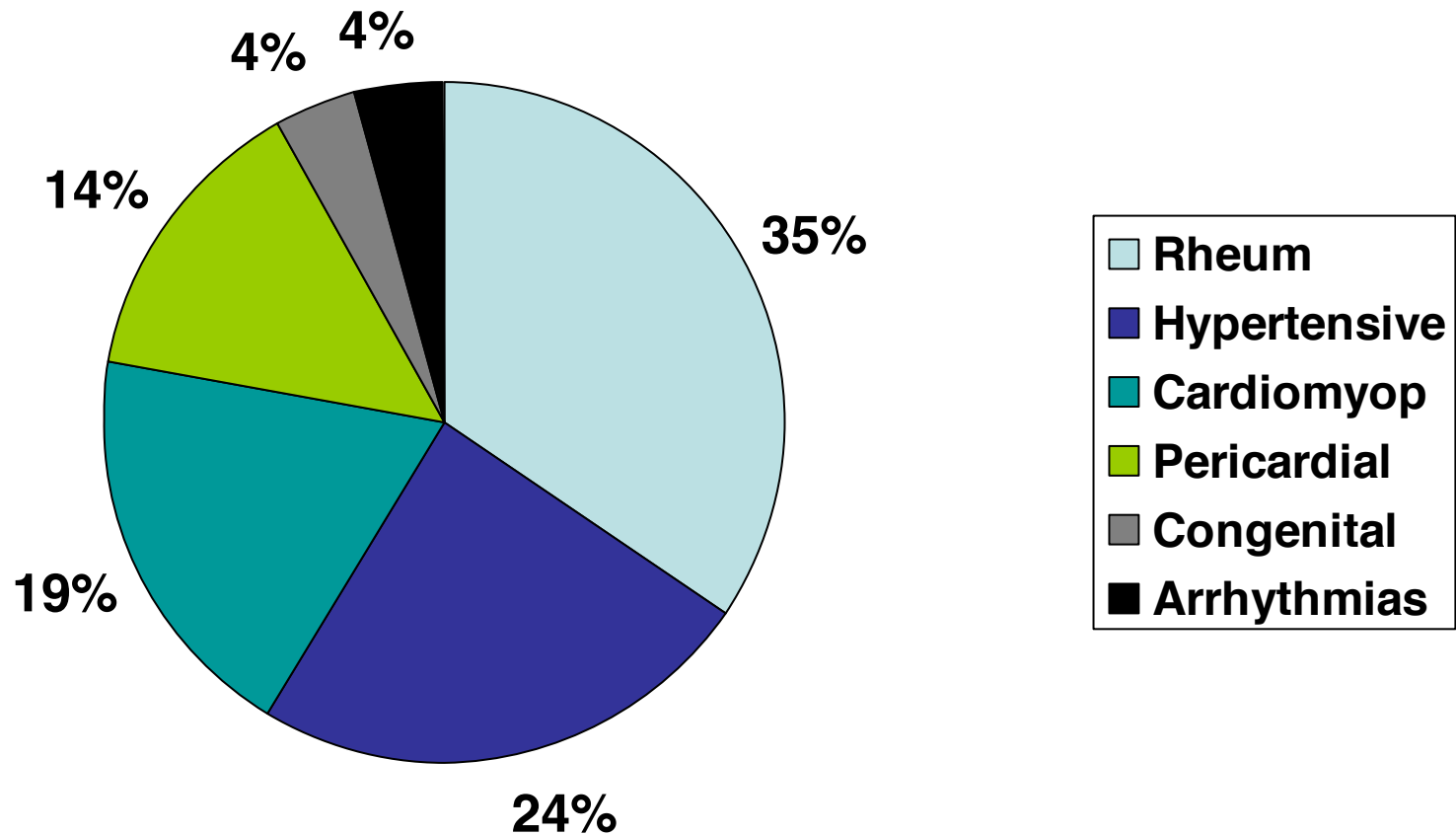
# Common Cardiac Disorders in Children in Malawi

Dr Neil Kennedy

# What causes heart disease in children in Africa?



# Heart disease in Mzuzu (adults + children 2001-6)



# Paediatric Cardiac Clinic QECH

- 47 (60.3%) have congenital heart disease (CHD). Of these, 31 have a left-to-right shunt (21 Ventricular septal defect , 6 Atrio-ventricular septal defect, 4 patent ductus arteriosus- PDA), 14 have cyanotic CHD (12 Tetralogy of Fallot , 2 complex cyanotic CHD) and 2 have pulmonary stenosis.
- 31 (39.7%) have acquired heart disease (20 rheumatic heart disease, 7 dilated cardiomyopathy, 2 Duchenne dystrophy, 2 endomyocardial fibrosis)

# What causes heart disease in children in Mozambique?

- 2170 school children in Maputo had echo
  - 5 (0.23%) congenital
  - 5 (0.23%) symptomatic rheumatic
  - 61 (2.8%) asymptomatic rheumatic

*Marijon et al NEJM 2007;375;470-6*

- 1030 rural sample adults + children had echo
  - 28% of 10-19 y.o. endomyocardial fibrosis
  - 7% symptomatic

*Mocumbi et al NEJM 2008;359:43-9*

# What will this talk be about?

- Presentation of heart disease in children
- Causes / treatment of failure:
  - Rheumatic heart disease
  - Congenital heart disease causing failure
  - Others: EMF / dilated cardiomyop
- Cyanotic Congenital heart disease
- Innocent / pathological murmurs

# How does Cardiac disease present in Children?

- CCF
- Chronic cough
- Cyanosis
- Malnutrition / failure to thrive
- Part of a syndrome eg Trisomy 21
- Isolated murmur
- Hypertension

## 3 *major* Cardiac Failure Signs in YOUNG Children

- TACHYCARDIA
- TACHYPNOEA
- HEPATOMEGALY
  
- Infants: poor feeding / sweating / poor growth
- Children:
  - R side: ↑ JVP, oedema
  - L side: Increased HR, RR, creps etc



# Causes of CCF in children

- Newborn:
  - Asphyxia
  - Sepsis
  - Congenital (PDA, TGA)
- 1/12 – 5 years:
  - **Anaemia**
  - **Congenital (VSD, PDA, AVSD)**
  - **Rheumatic – acute / chronic**
  - Sepsis / pneumonia
  - Pericarditis
  - Cardiomyopathy
- 5 +:
  - **Rheumatic – acute / chronic**
  - **Cardiomyopathy**
  - Pericarditis
  - Hypertension with GN

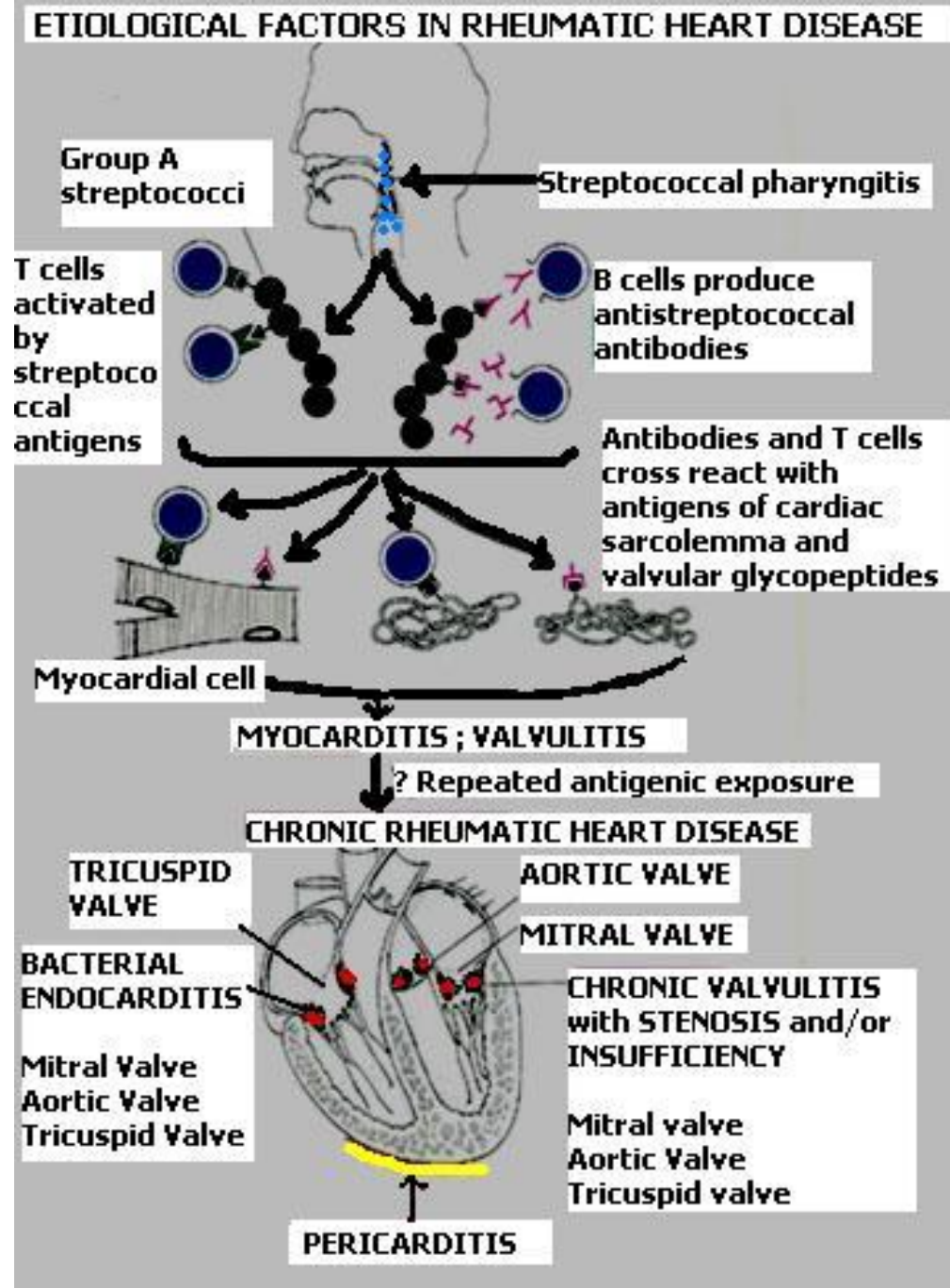


# Rheumatic Fever - epidemiology

- 16-20 million people affected worldwide
- 250,000 deaths / year worldwide
- Prevalence: 10-15/1000 SSA
- Associated with poverty / urban slums / LBW / overcrowding
- Mainly children 6-10 years (starts 3+)
- Reducing incidence industrialised nations

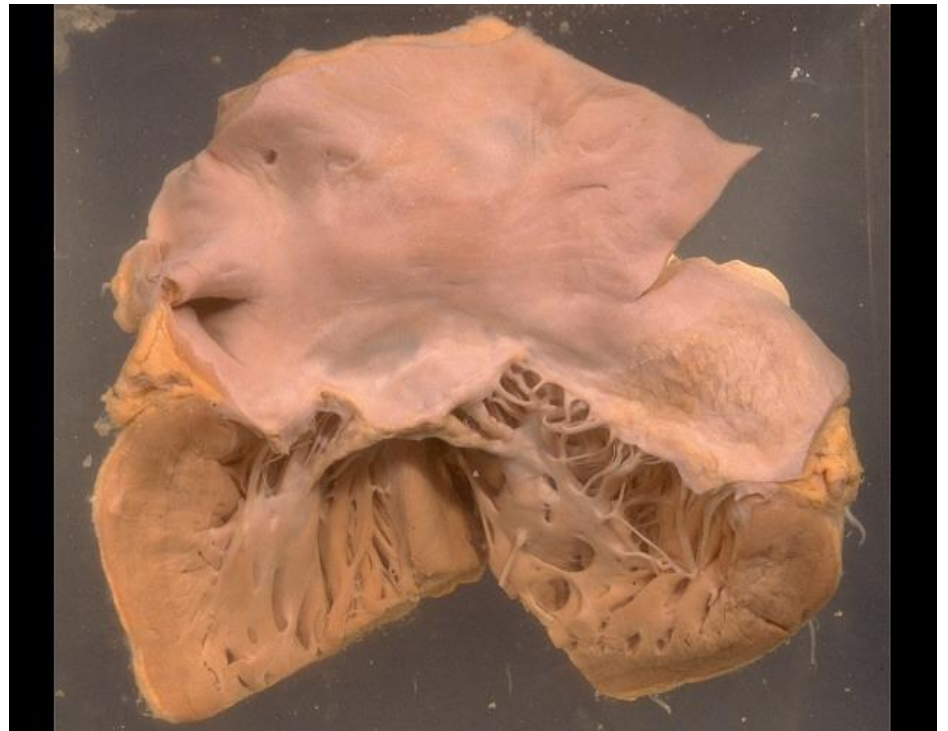
# Rheumatic fever - pathogenesis

- 0.3 - 3% of cases of GA $\beta$ HS (*Strep pyogenes*)
- 1-5 week incubation
- 'Molecular mimicry'
- MBL2, TNFA linkage
- M protein
- Heart, joints, brain, connective tissue involved
- Low IL4 in valvular tissue allows chronic inflammation



# Rheumatic fever - pathology

- Pancarditis
- Endocarditis esp. L heart
- Oedematous → thickened, scarred MV
- Shortened papillary muscles preventing valve apposition
- Aschoff Nodules
- Jt effusion



# RF – Clinical Features

- FEVER often  $> 39$ , may  $\downarrow$  but persist for 3 weeks
- Arthritis – 75%:
  - Large joints, beginning legs. Migrating
  - Asymmetrical, painful ++
- Carditis – 50%:
  - New murmur (MR++),  $\uparrow$ HR, CCF, dyspnoea
  - Sometimes pericarditis, chest pain
- Chorea – 20% (Sydenhams)
- Erythema Marginatum – 10% (often missed)
- Subcutaneous nodules – 5% (extensor surfaces)



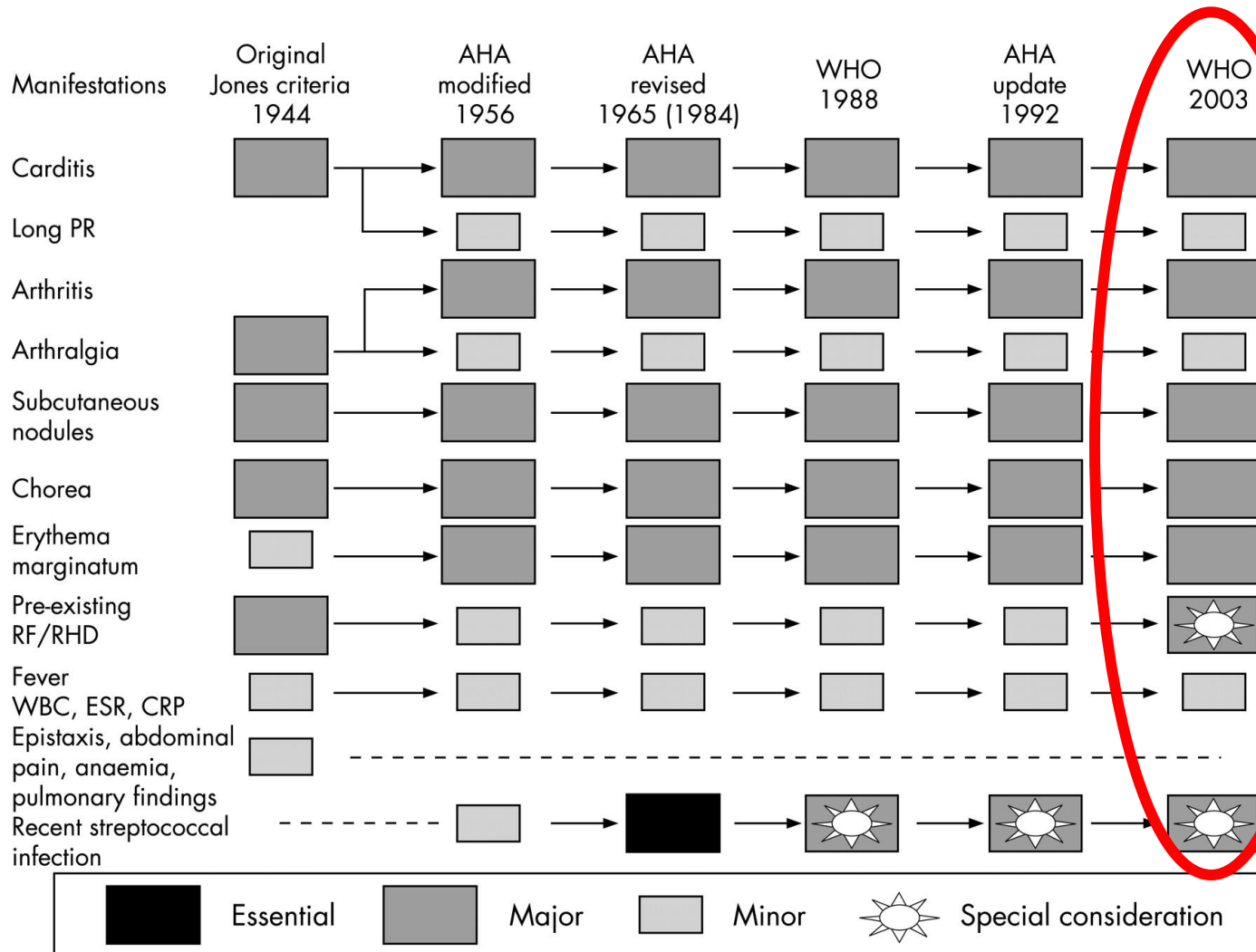


# RF – Diagnosis – Jones Criteria

- Evidence of recent Strep:
  - ASOT > 200, +ve culture, scarlet fever
- 2 major *or* 1 major + 2 minor
- Major:
  - Arthritis, carditis, nodules, chorea, erythema marg., pre-existing RF
- Minor:
  - Fever, ↑ WCC / CRP / ESR, ↑PR interval



# Changes in Jones Criteria



Nkomo, V. T Heart 2007;93:1510-1519

# Acute RF - Treatment

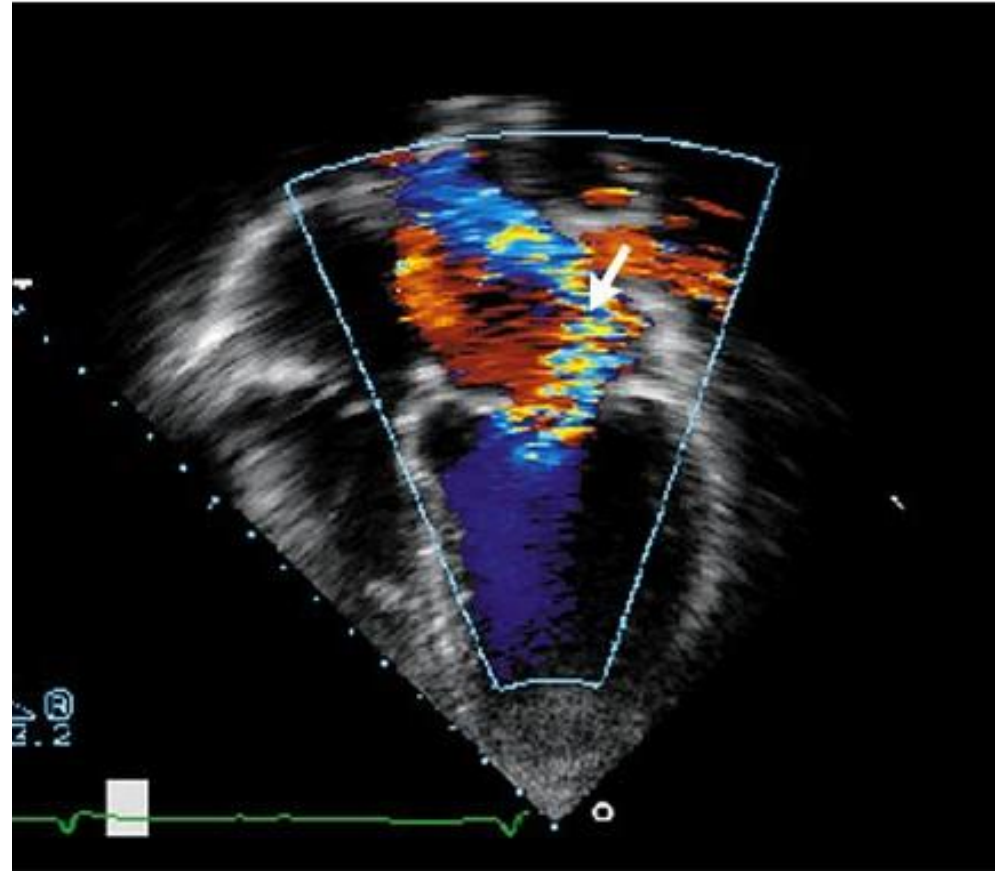
- Bed rest
- Kill strep – penicillin x 10/7
- Aspirin 25mg/kg QDS until no inflammation (expect dramatic response)
- Prednisolone if carditis
- Treat failure
  
- **~ 75% will progress to Chronic RHD**
  - **Repeated episodes / poor**

**GET THEM ON  
MONTHLY  
BENZATHINE**

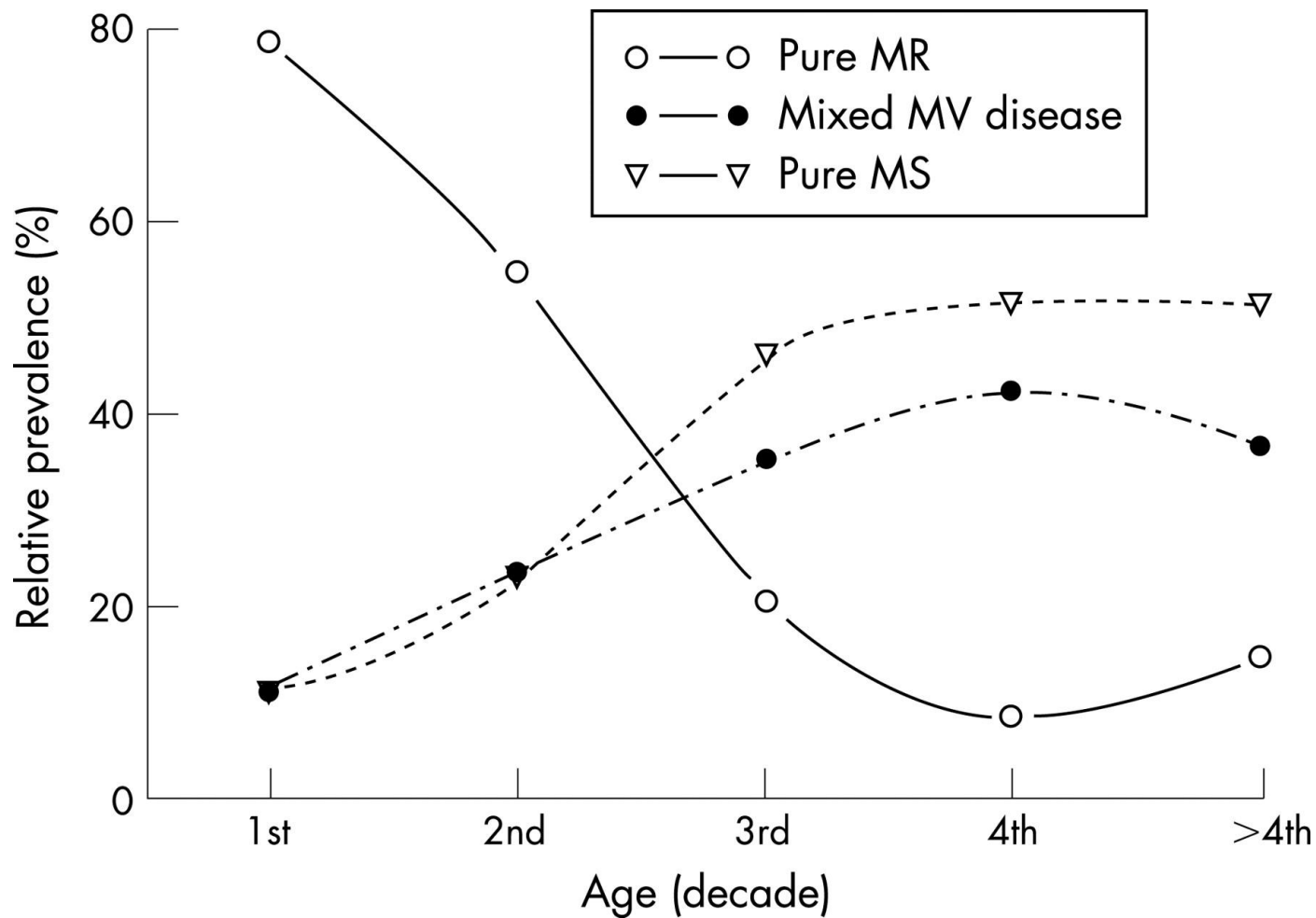
# Chronic RHD

– valves involved in children

- 85% Mitral:
  - 65% MR only
  - 17% MR + MS
  - 3% MS only
- 8% Aortic
- 6% Tricuspid



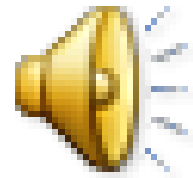
# Increasing MS with time



Nkomo, V. T Heart 2007;93:1510-1519

# Chronic RHD - clinical

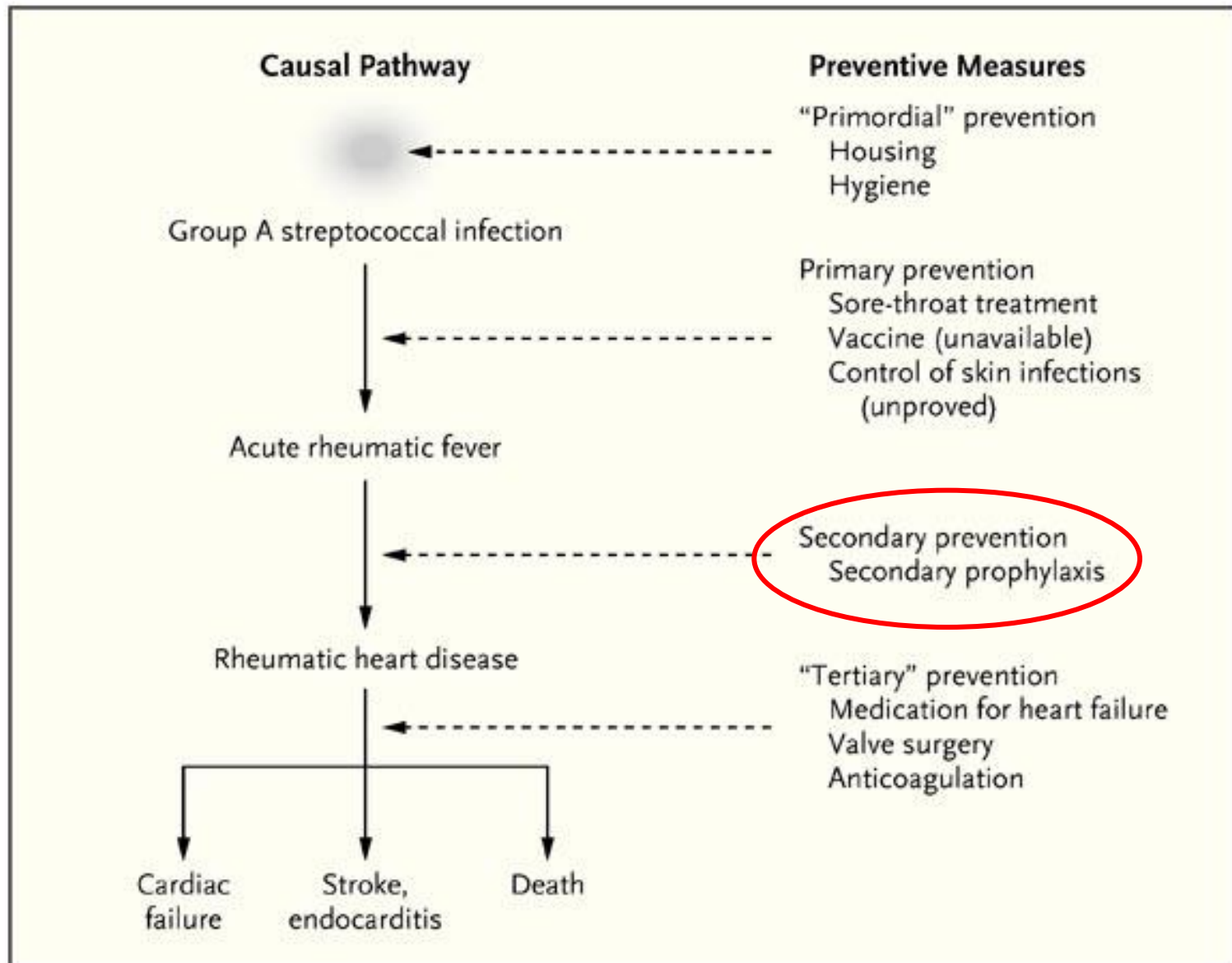
- CCF
- Diffuse heaving apex
- Systolic thrill
- Blowing MR murmur at Apex
- EDM of MS (turn on L)
- Aortic regurg diastolic murmur
- CXR: cardiomegaly, ↑LA, CCF
- ECG: LV hypertrophy



# Chronic RHD - treatment

- Secondary prevention:
    - Monthly benzathine penicillin
  - Surgery if available
  - Treat CCF:
    - frusemide
    - digoxin
    - ACE inhibitors
    - $\beta$  Blockers
- } Extrapolation from adult studies only

# Prevention of RHD





**GET THEM ON  
MONTHLY  
BENZATHINE**

# Congenital heart disease

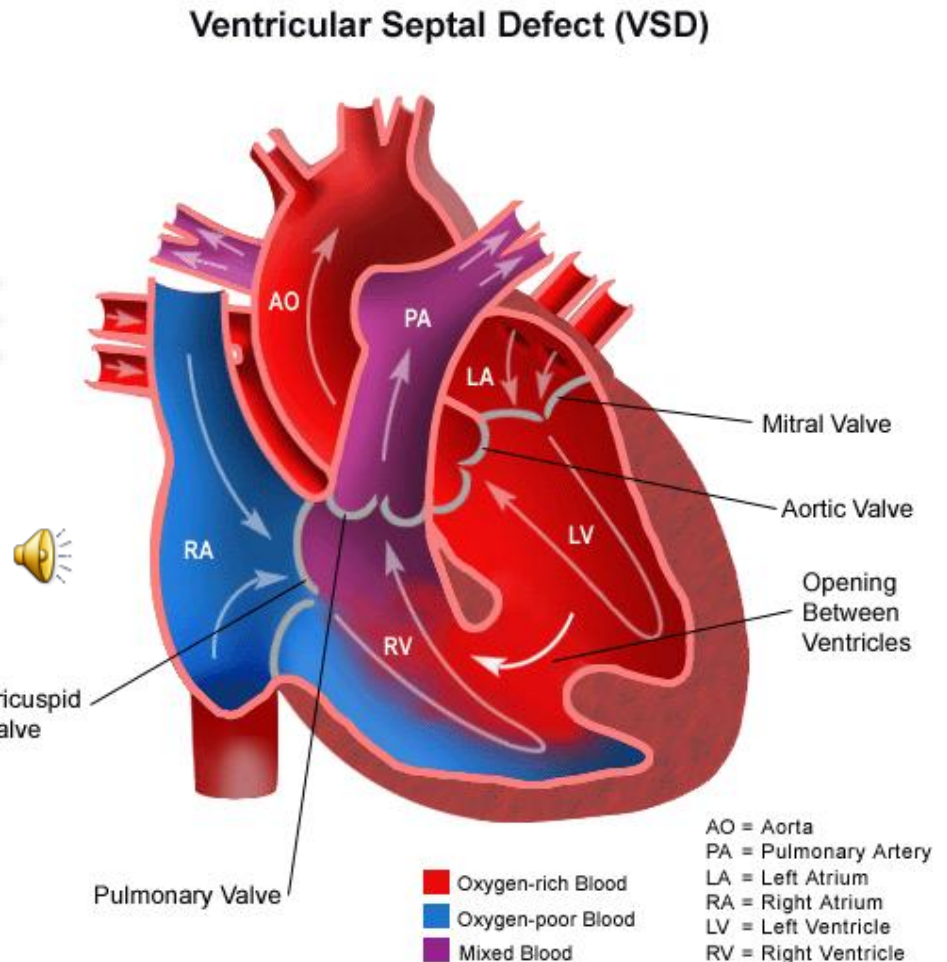
- 1% of population
- Presents as:
  - CCF
  - Cyanosis
  - Chronic cough
  - Syndrome
  - Hypertension (Coarctation)
  - FTT

# Congenital HD Causing CCF

- Usually L → R shunt:
  - Ventricular Septal Defect
  - AVSD (less often ASD)
  - Patent Ductus Arteriosus
- Obstructive lesion:
  - Pulm stenosis

# Ventricular septal defect

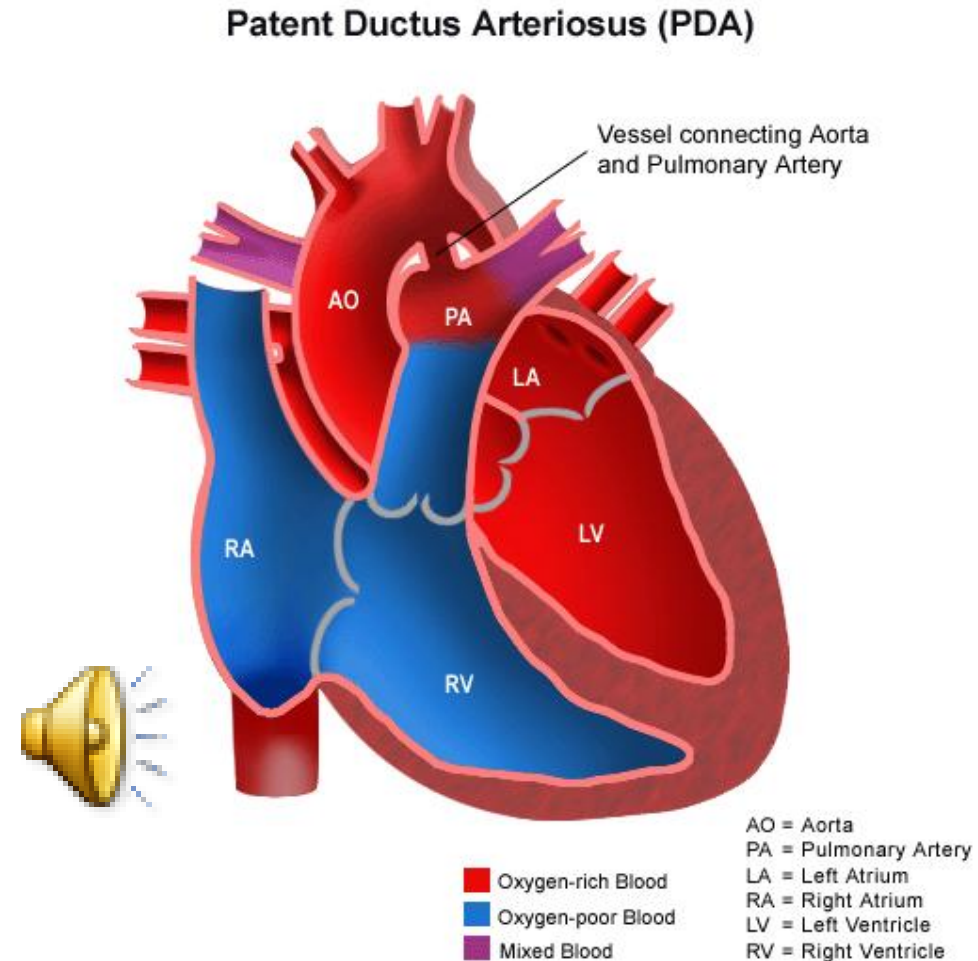
- Commonest CHD
- L to R shunt
- Harsh PSM at LSE and apex
- Small holes close
- Big holes cause:
  - CCF
  - Shunt reversal (Eisenmenger's)



# VSD Video

# Patent Ductus Arteriosus

- Failure to close after birth
- L to R shunt
- Premature babies++
- Systolic / machinery murmur
- Bounding pulses
- Causes CCF
- Surgery in Malawi



# Other causes of CCF

- Dilated cardiomyopathy:
  - post-viral – Coxsackie / HIV
  - Important cause in children over 5
- Endomyocardial fibrosis:
  - Hot and humid tropics (Uganda ++, increasingly recognised in Malawi)
  - (5-20) years
  - Aetiology unknown
  - Endomyocardial fibrosis R > L → TR / MR
  - Ascites ++ (as well as other signs R sided / CCF)
  - Eosinophilia





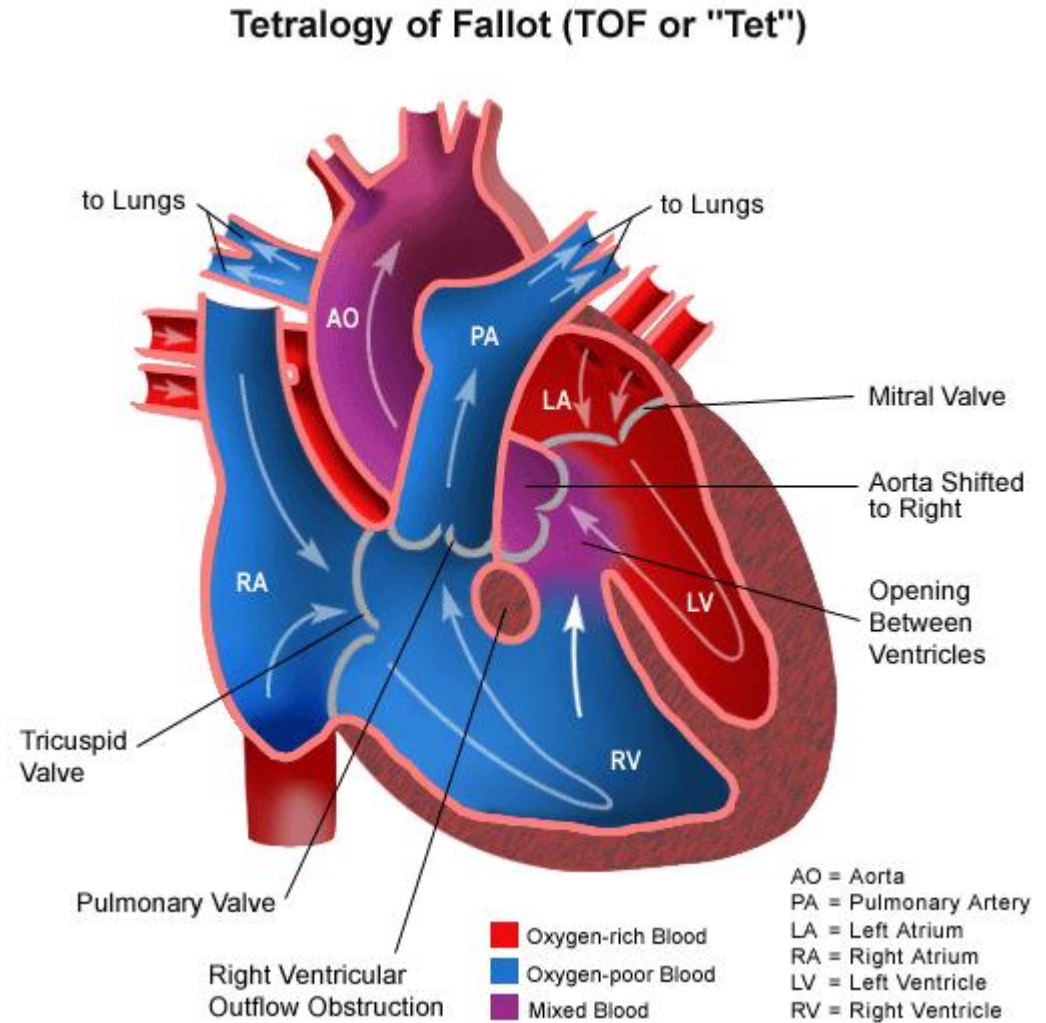
# Cyanotic Congenital HD

- R → L shunt
- Cyanosis OFTEN missed
- Differentiate from resp cyanosis by:
  - Nitrogen washout (oxygen)
- Soon after birth:
  - Transposition Great Arteries (TGA)
- Infancy / childhood:
  - Tetralogy of Fallot (TOF)
  - Cyanotic 'spells'



# Tetralogy of Fallot

- Consists of:
  - VSD
  - Pulmonary stenosis
  - RVH
  - Over-riding aorta
- Treat surgically (RSA)
- Medical Rx:
  - propranolol



# Innocent vs non-innocent murmurs

- 30-40% of ALL children have a murmur at some time
- Most are innocent
- ↑ after exercise / fever / anaemia
- Innocent murmurs are:
  - Soft / systolic / short
  - Asymptomatic
  - Localised
  - Vary with posture
  - Have normal CXR / ECG

Pali mafunso?



# You need to know....

- How heart disease presents in children
- Common causes of CCF:
  - Rheumatic fever / heart disease
  - Congenital HD causing CCF
- Treatment of CCF in children
- Common causes of cyanotic congen HD:
  - Tetralogy of Fallot
- Features of an innocent murmur