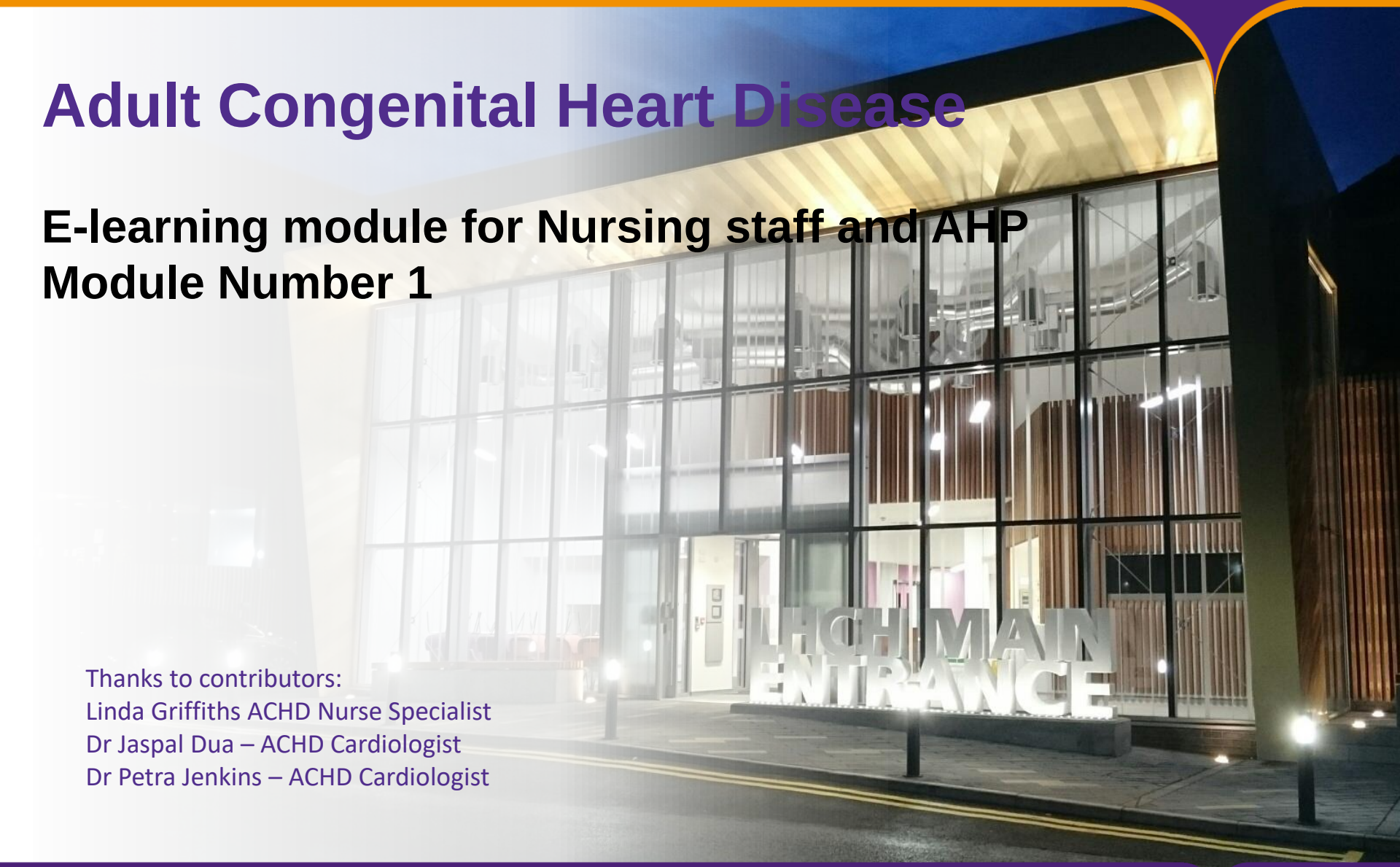
Adult Congenital Heart Disease

E-learning module for Nursing staff and AHP
Module Number 1



Thanks to contributors:
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Dr Jaspal Dua – ACHD Cardiologist
Dr Petra Jenkins – ACHD Cardiologist

- Welcome to this e-learning module. This is a brief introduction to adult congenital heart disease in the North West of England, Isle of Man and North Wales.

- This is not intended to be exhaustive and therefore we would recommend that you invest time in self learning

By the end of module one we would expect you to have/know:

- A brief understanding of why we need a specialist service for patients with ACHD
- Know what the new service model looks like in the North West of England, North Wales and Isle of Man
- An appreciation of the need for good transition to adult services
- An overview of common ACHD defects

Background

- The number of adults with Congenital Heart Disease has overtaken the number of children
- This new population of patients with Congenital Heart Disease requires a specialist service to meet their unique needs.
- Catchment population for the North West estimated >7,500,000
- In 2006 a health needs assessment estimated the ACHD prevalence to be 23,701 patients by 2010 – this was expected to rise 10% year on year
- Most of these patients are not known to the service

Background

- It’s a dramatic success story of modern medicine
- Its created a new population of patients who have not been cured
- Corrective surgery is a myth for many
- Many face prospect of further surgery, arrhythmias and heart failure and many are at risk of sudden death

NHS England Standards

- In September 2015 NHS England published national standards by which congenital heart disease services were to be measured in England.
- NHS England announced their decision on 30th November 2017.
- Please click the link below to see the standards that we need to meet
- https://www.england.nhs.uk/commissioning/wp-content/uploads/sites/12/2016/03/achd-l1-standards-2016.pdf

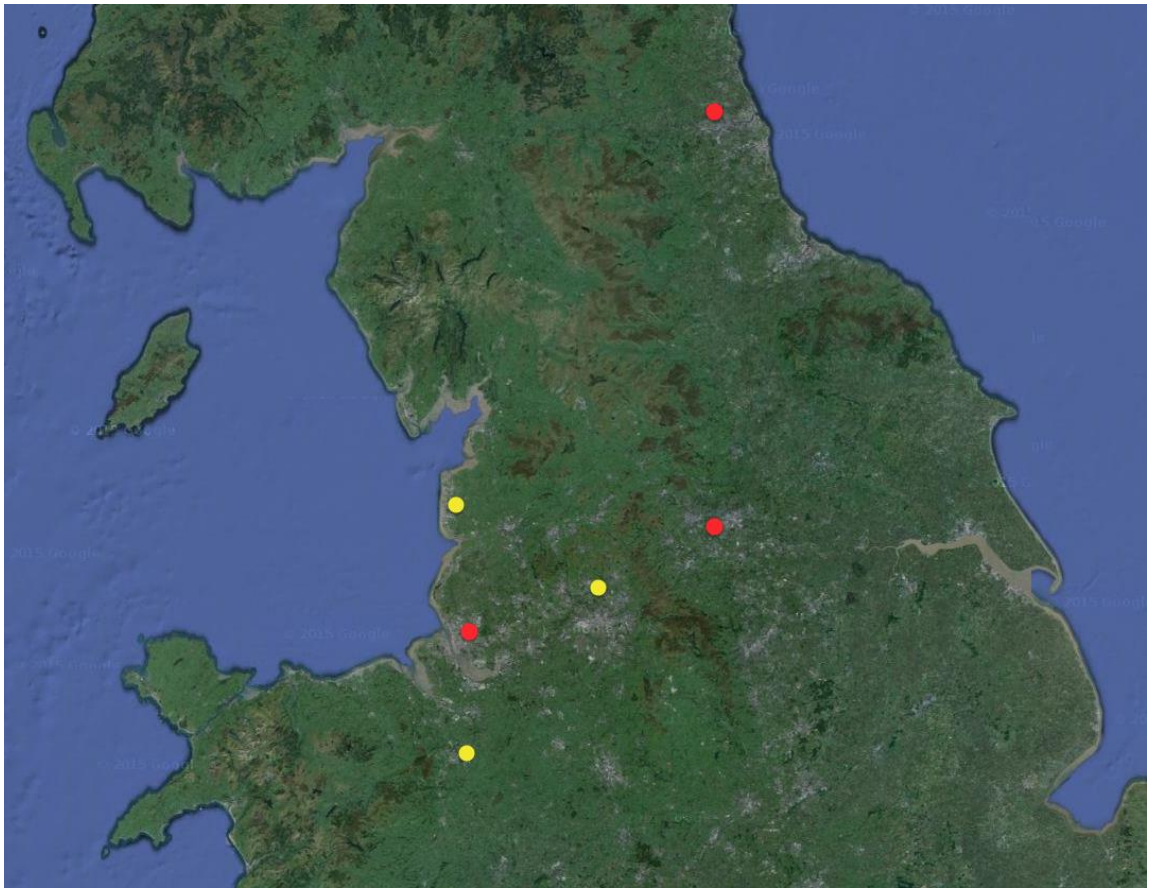
ACHD Model of Care for the North West

- **Level 1 Surgical Centre**
- **Level 2/3 Centres**

Level 1 ACHD Centre:
Liverpool Partnership: Liverpool Heart + Chest Hospital, The Royal Liverpool Hospital and The Women's Hospital

Level 2 ACHD Centre:
Manchester Royal Infirmary

Level 3 ACHD Centre
Wrexham Maelor



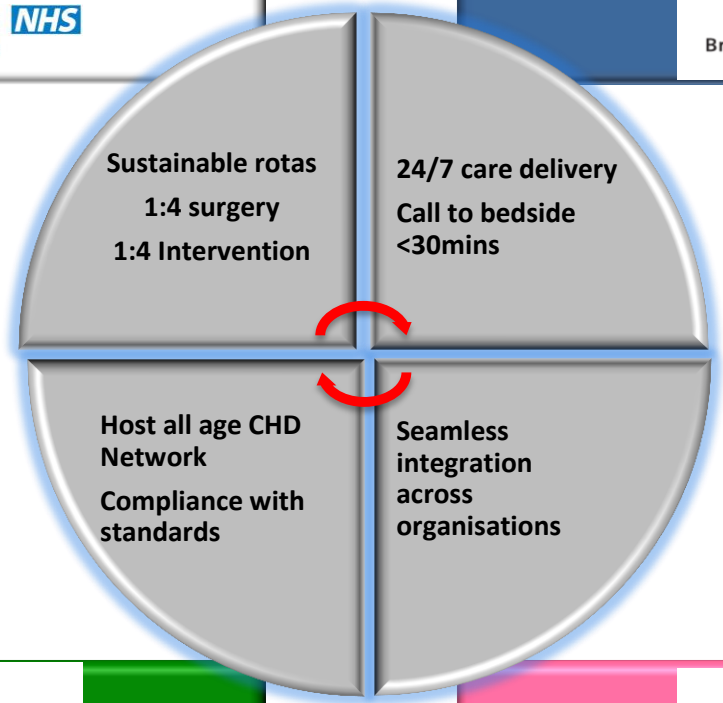
New North West Model of Care for ACHD
The North West Liverpool Partnership

Liverpool Heart and Chest Hospital NHS
NHS Foundation Trust

- Adult Level 1 surgical centre
- Adult interventions
- Shared Adult Services with Royal
- Transition services with Alder Hey

The Royal Liverpool and Broadgreen University Hospitals NHS
NHS Trust

- Medical/cardiac admissions for ACHD
- Non- cardiac surgery for ACHD
- Delivery of high-risk cardiac patients with LWH
- Provide adult co-dependencies for LHCH
- ACHD cardiology on call



Alder Hey Children's NHS
NHS Foundation Trust

- Paediatric Level 1 Surgical Centre
- Paediatric co-dependencies
- Common surgical rota with LHCH
- Support LHCH anaesthesia
- Surgery for neonates with CHD together with LWH

Liverpool Women's
NHS Foundation Trust

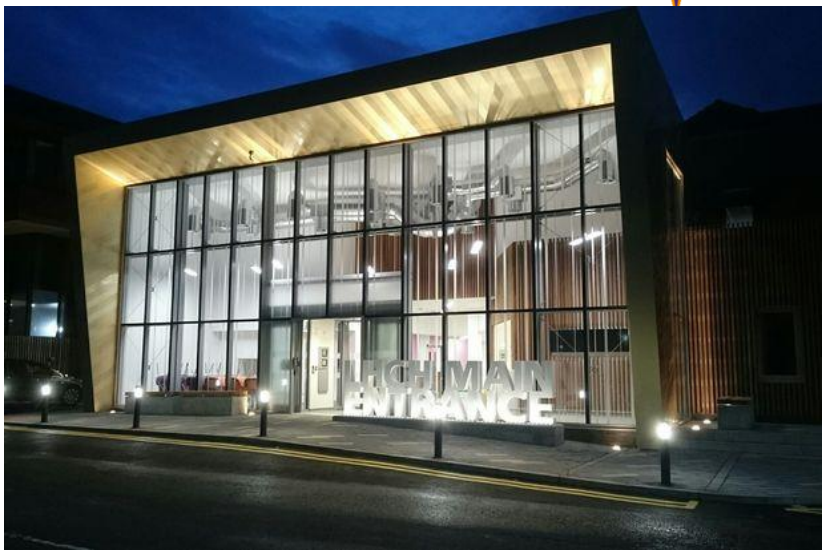
- Manage high risk deliveries at Royal
- Pre pregnancy counseling
- Foetal Screening
- Coordinate delivery/intervention for neonates with CHD

North West CHD Partnership
A Lifetime in CHD Care

LIVERPOOL HEALTH PARTNERS

What are we hoping to achieve

- Provide specialist CHD care 24/7
- Sustainable on call rotas
- Surgery/Intervention performed in high volume centres by highly experienced operators
- Achieve world leading outcomes
- Deliver an all age CHD Network across the North West
- Work with providers across the North West



Transition

- About 400 young people will be transferring their care to adults each year
- They will have experienced a very patient and family centred, high quality service throughout their childhood
- They will come with fears, anxieties, past experiences that will influence how they cope in the adult setting
- It is important that as an adult practitioner you appreciate the issues facing young people

Where Do They Fit In?

Paediatric Setting

Infantilised & dependent

Family centred

Developmentally focused

Ignores growing independence & adult behaviour

Adult Setting

Medically centred

Acknowledges patient autonomy

Reproduction & employment

Neglects growth & development & family concerns

Neither paediatric or adult staff are trained in adolescent health care!!

https://youtu.be/Q_cTv-1eOEg

Overview

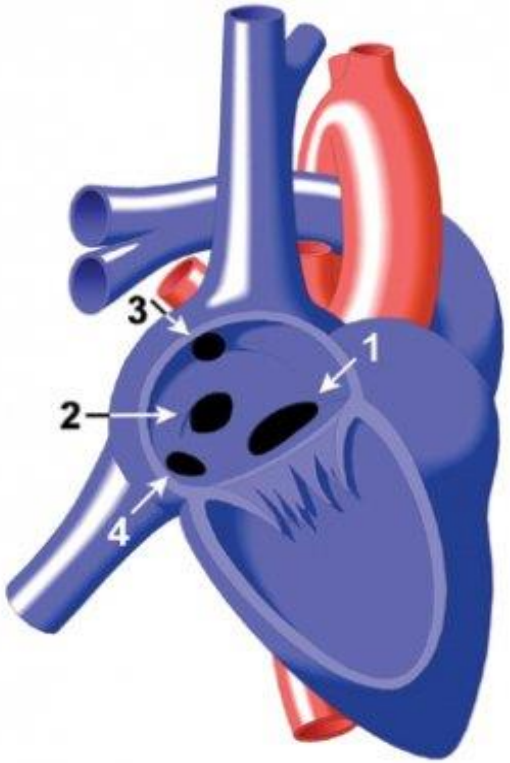
- The next slides will introduce you to a number of congenital heart defects.

- Please take the time to listen to each pod cast

- **Acknowledgments:**
  You tube pod casts delivered by the ISACHD teaching course on Congenital Heart Disease
  http://achdlearningcenter.org/category/learning-modules/

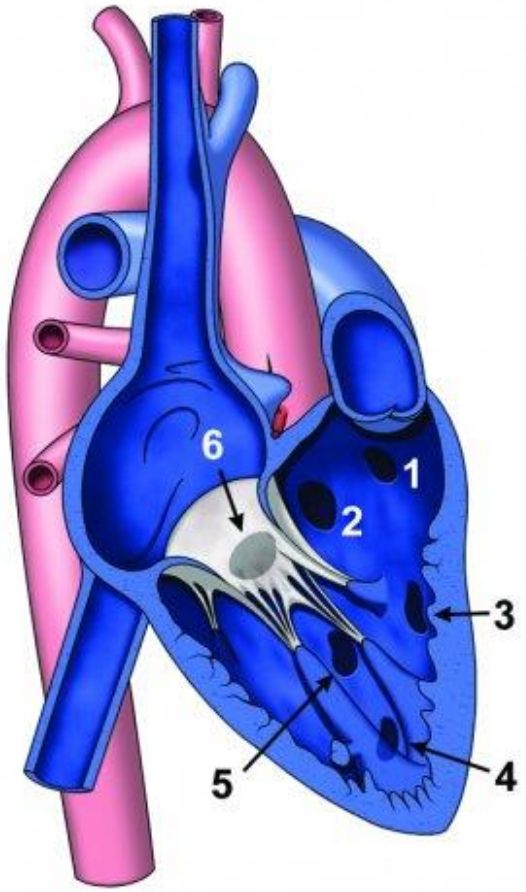
  Pictures from www.pted.com

Atrial Septal Defects



1. Primum, 2 - Secundum
3 - Superior Sinus Venosus
4 - Inferior Sinus Venosus

- ASDs vary in size and position within the atrial septum
- ASD's vary in the severity of symptoms they may cause
-
- They account for between 6 to 10% of all cases of congenital heart disease
- twice as prevalent among girls as boys.
- https://youtu.be/-fuQZ2J2pTc



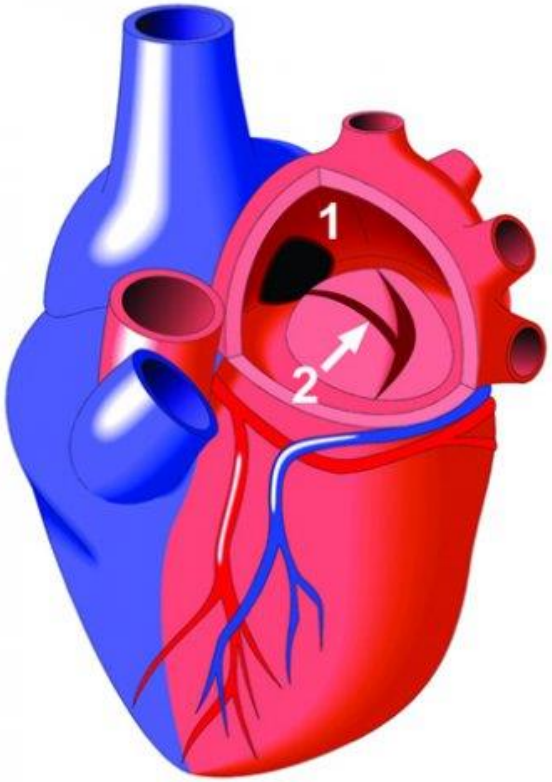
Types of VSD

1. Outlet
2. Perimembranous
3. Muscular
4. Apical
5. Mid-muscular
6. inlet

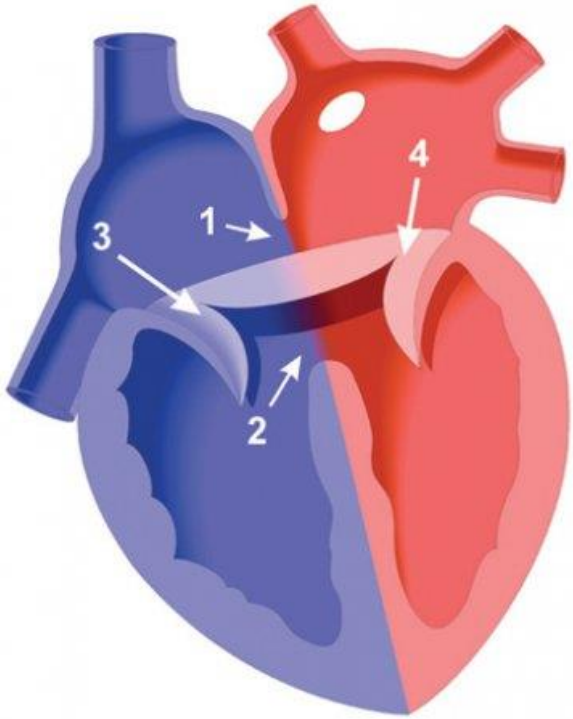
https://youtu.be/fOpTkoye6nw

- 90% Partial AVSD's occur in non Down's Syndrome patients
- 75% Complete AVSD's occur in Downs Syndrome patients
- AVSD may also occur in association with Tetralogy of Fallot and other forms of complex congenital heart disease

Atrio-Ventricular Defect



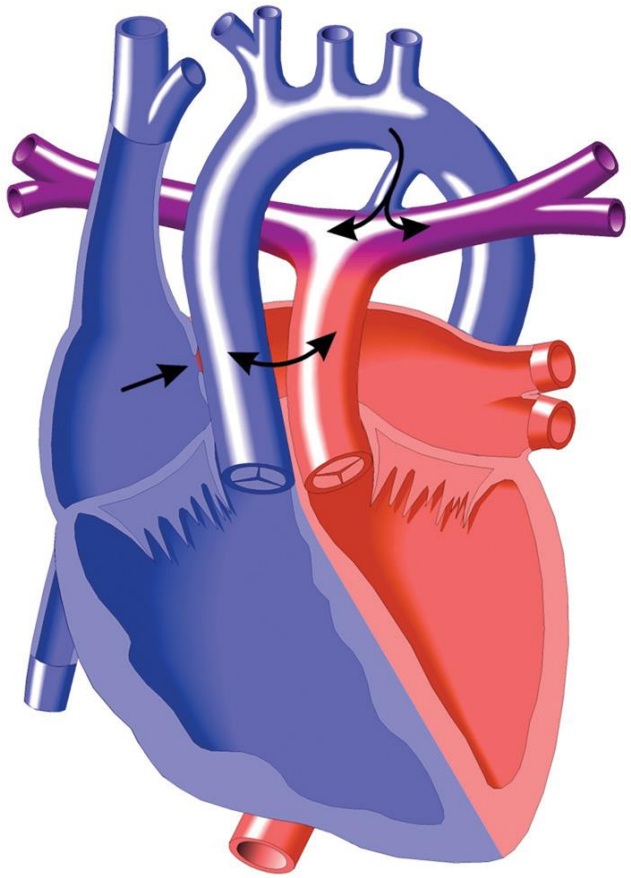
Partial AVSD with cleft in the mitral valve



Complete Atrio-Ventricular Septal Defect

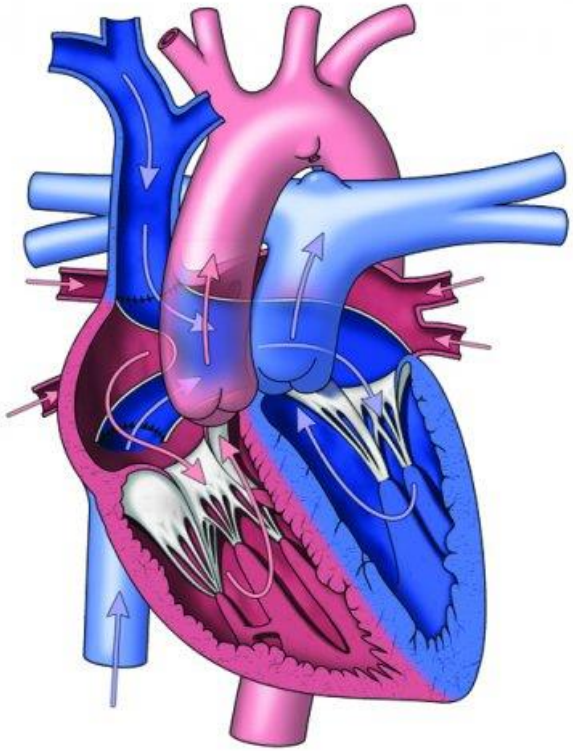
https://youtu.be/cpQNkg2yfYU

Simple Transposition of the Great Arteries

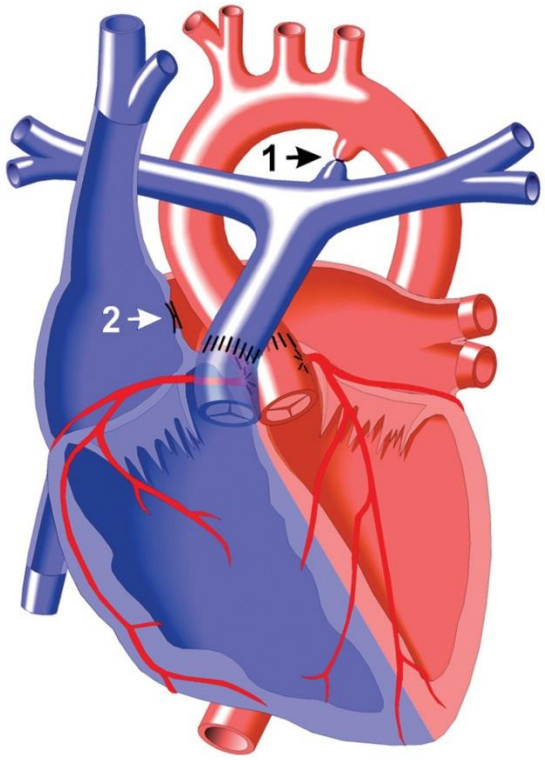


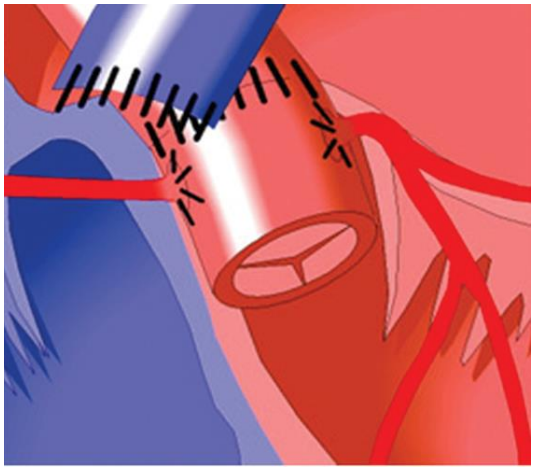
- Survival is dependent upon mixing of blood
- Mixing occurs initially via PFO and PDA
- Is incompatible with life 90% die before age 1 yr
- Therefore an adult will have had some form of surgical palliation

Surgical Procedures for TGA



Mustard/Senning Procedure





Arterial Switch Procedure

https://youtu.be/4U4vCIOGohg

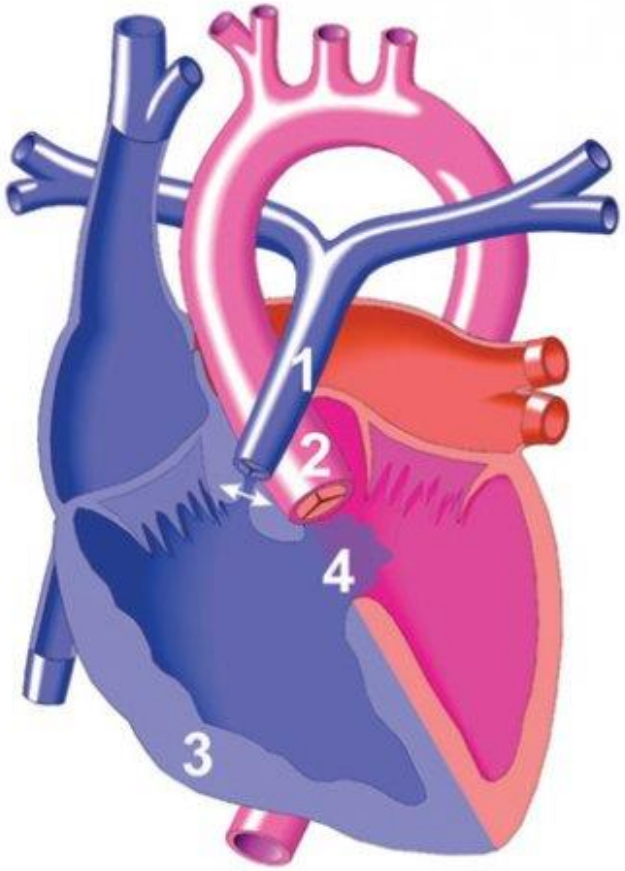
https://youtu.be/cozGGXcS_n8

One of the most common forms of cyanotic congenital heart disease

Accounts for up to 10% of all forms of CHD

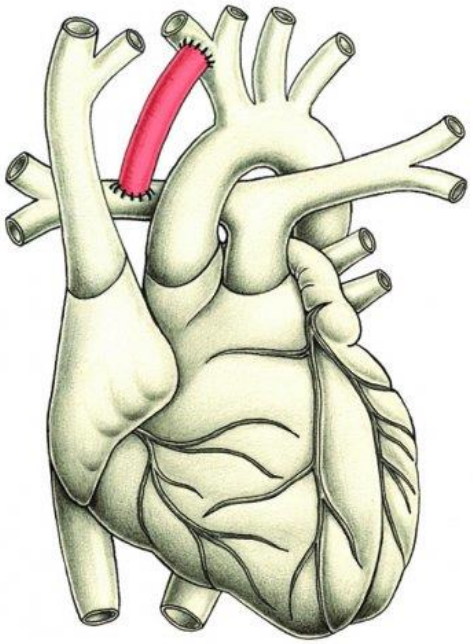
Approximately 15% of patients with Tetralogy of Fallot have deletion of chromosome 22q11 (Di-George Syndrome)

Tetralogy of Fallot

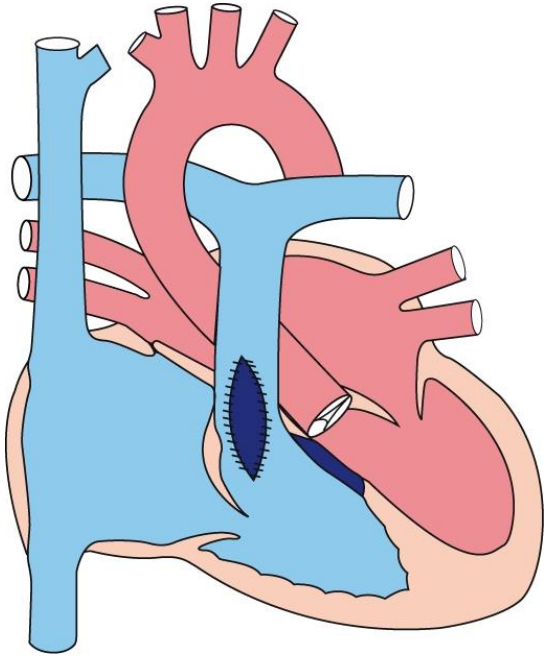


- A large ventricular septal defect (VSD)
- An enlarged aorta which overrides the VSD
- hypertrophy (enlargement) of the right ventricle
- constricted and obstructed outflow from the right ventricle - pulmonary stenosis and infundibular stenosis

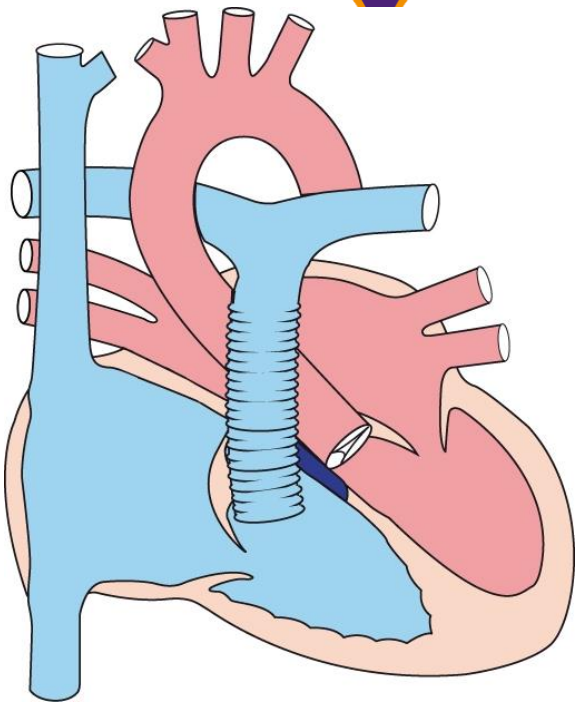
Surgical procedures for Tetralogy of Fallot



Blalock Taussig Shunt (modified)



Transannular patch repair



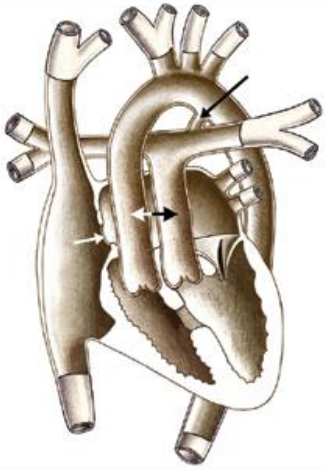
Right ventricular to pulmonary artery conduit/homograft

https://youtu.be/ra7n69y0eOs

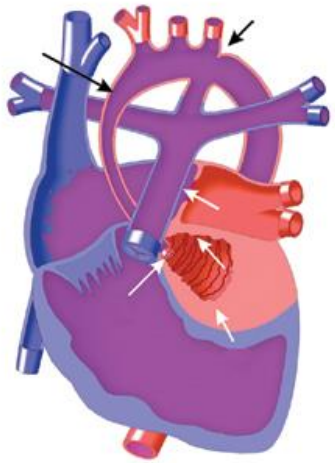
The single ventricle

- Accounts for less than 1% of congenital defects
- Small numbers but the most complex
- Truly solitary ventricles are exceedingly rare
- Mostly there is a second rudimentary or hypoplastic ventricle present
- “Functionally single ventricle”

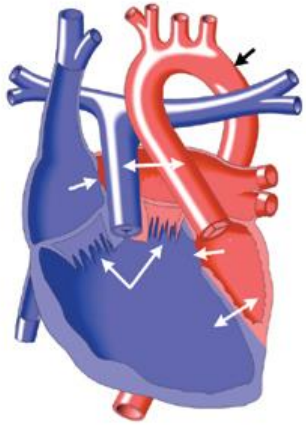
Types of Single Ventricle Anatomy



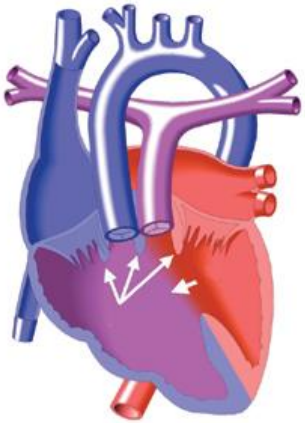
Tricuspid Atresia



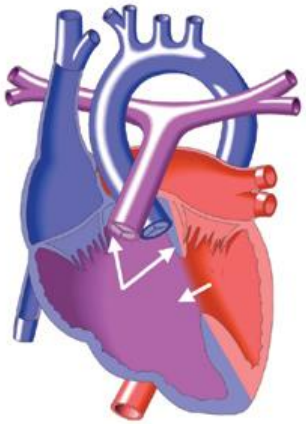
Hypoplastic Left Heart Syndrome



Double Inlet Left Ventricle

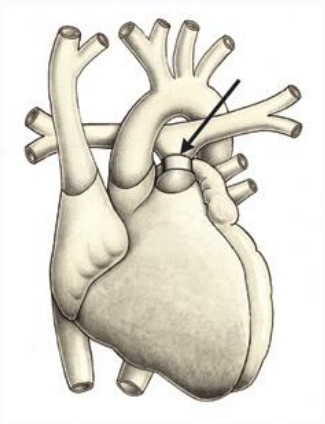


Double Outlet Right Ventricle w/Transposition

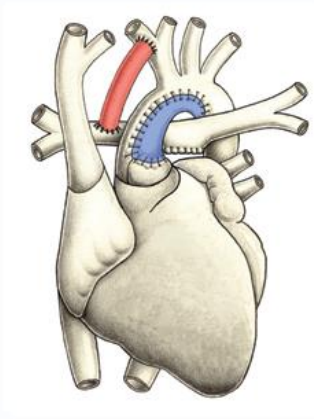


Double Outlet Right Ventricle

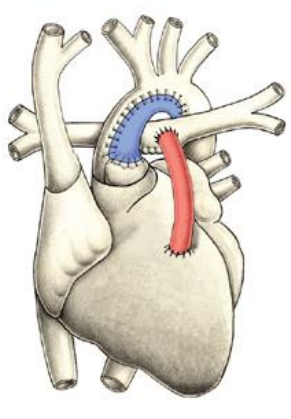
Surgical procedures for single ventricle



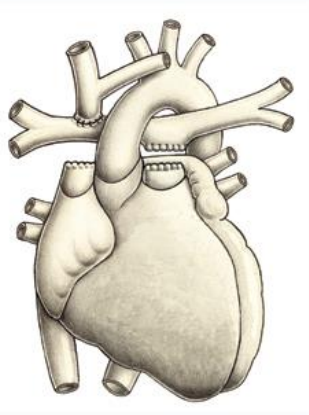
Pulmonary Band



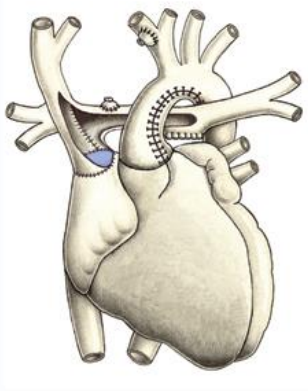
Norwood Procedure



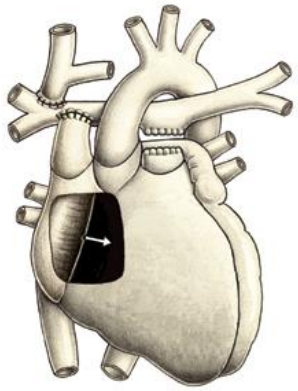
Sano Modification of the Norwood



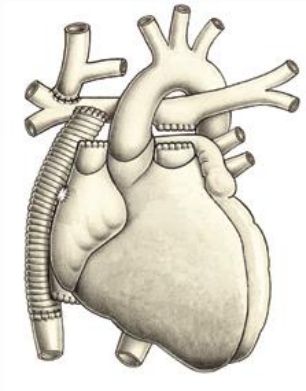
Glenn Shunt Procedure



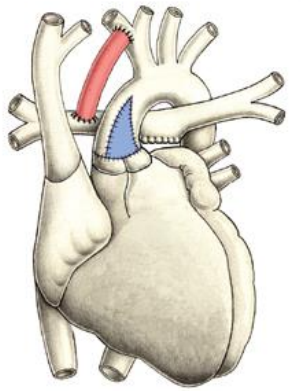
Hemi-Fontan Procedure



Fontan Procedure (Lateral Tunnel)



Fontan Procedure (Extracardiac)



Damus-Kaye-Stansel Procedure

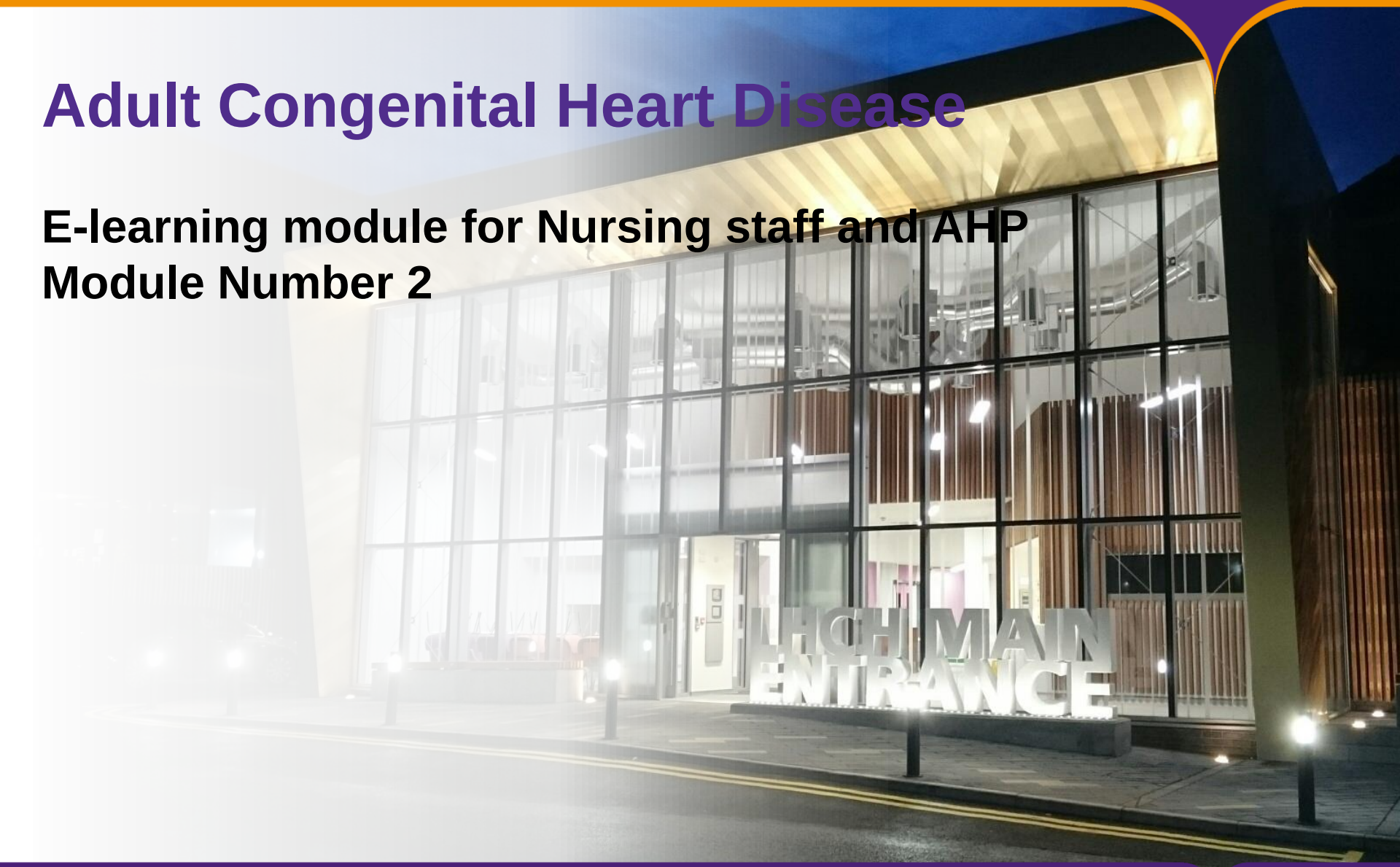
https://youtu.be/GyVcvMc29KA

- Arrhythmias
- Thrombo-embolic events
- Fontan obstruction
- Heart failure
- Hepatic problems
- Protein losing enteropathy
- Cyanosis

https://youtu.be/Cj-ISt1teIM

Adult Congenital Heart Disease

E-learning module for Nursing staff and AHP
Module Number 2



By the end of module two we would expect you to have/know:

- An understanding of common ACHD emergencies and how to treat them
- Principles of care unique to ACHD
- The Mental Capacity Act and how it relates to ACHD
- How to contact the ACHD Team

Common Emergency Presentations

- Arrhythmias
- Chest pain
- Blue patients
  - General acute deterioration
  - Haemoptysis
  - Cerebral abscess / Stroke
  - Cholecystitis
- Endocarditis
- Coarctation repair site problems
- Thrombosed prosthetic valves
- Non-cardiac emergencies

- DON'T PANIC!
- **Initial** treatment just the same as any other unwell patient
- Assess Airway Breathing Circulation (BLS)
- Manage arrhythmias as arrhythmias
- Manage heart failure as heart failure
- Manage chest pain as chest pain

In majority

- Commonest presenting complaint in local A&E by far
- Manage as you would any other adult patient with an arrhythmia, just with a little more respect - Patients may deteriorate more quickly than you expect
- If compromised; need to be cardioverted electrically
- If not compromised; seek senior / medic advice
- DO NOT send them home (TRIAGE)

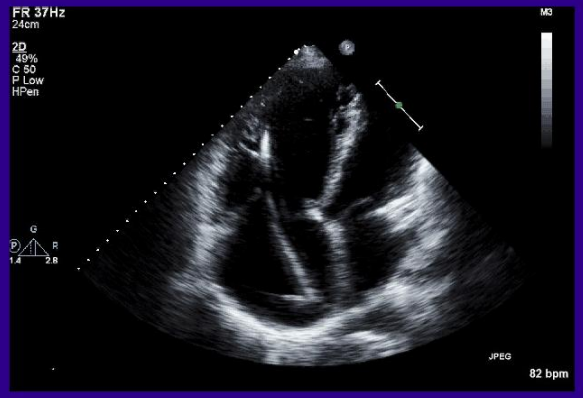
Arrhythmia

- Common in any operated ACHD patient
- Related to atrial scars, atrial volume or pressure load
- Atrial flutter (typical or atypical) most common, AF, re-entrant tachycardias less common
- Lead to syncope (You may make this problem worse by giving IV amiodarone):
- Particularly common in tetralogy of Fallot, atrial surgery, Mustard/Senning, Fontan, Ebsteins anomaly.

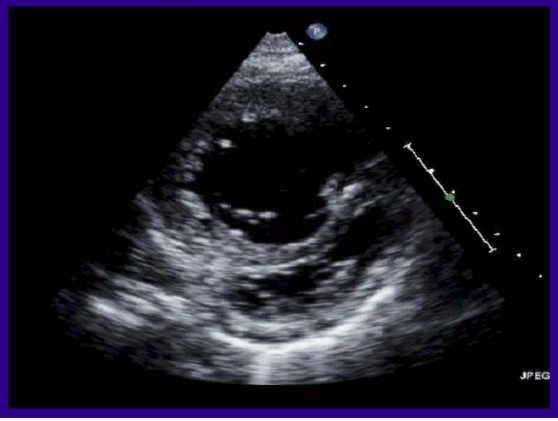
https://youtu.be/n7ImoVjSASI

Ventricular Tachycardia

VT is common in repaired tetralogy of Fallot with dilated RV

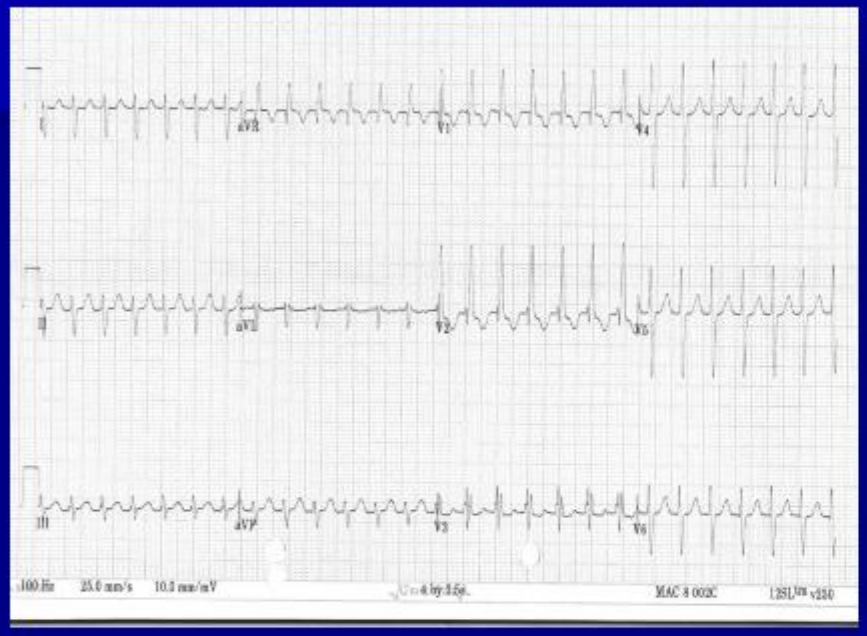


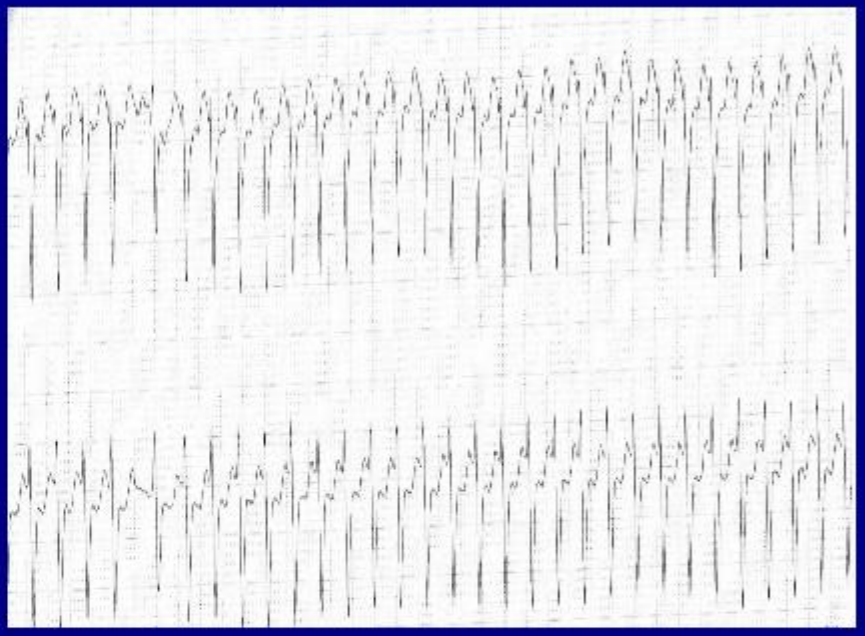
VT is common in Mustard/ Senning / ccTGA patients with failing systemic RV



https://youtu.be/EUHrgViYOsY

IV Amiodarone





Atrial flutter at about 300 / minute with 2:1 block

Ventricular response rate 150 bpm

Patient stable

Atrial flutter at about 250 / minute with 1:1 conduction

Ventricular response rate 250 bpm

Patient unstable

How to cardiovert a Fontan

- IV maintenance fluids to maintain filling pressures
- Explain the physiology in detail to the anaesthetist
- External pads on in case need to pace them afterwards
- Avoid prolonged positive pressure ventilation

How not to cardiovert a Fontan

Admitted at 5 pm in flutter

↓

Put on morning list for DCCV

↓

NBM overnight, no IV fluids

↓

Cardioversion delayed because of an emergency case

↓

At induction of anaesthesia
Low filling pressures because of dehydration
Vasodilatory anaesthetic agents
IPPV

↓

Avoidable circulatory collapse and death

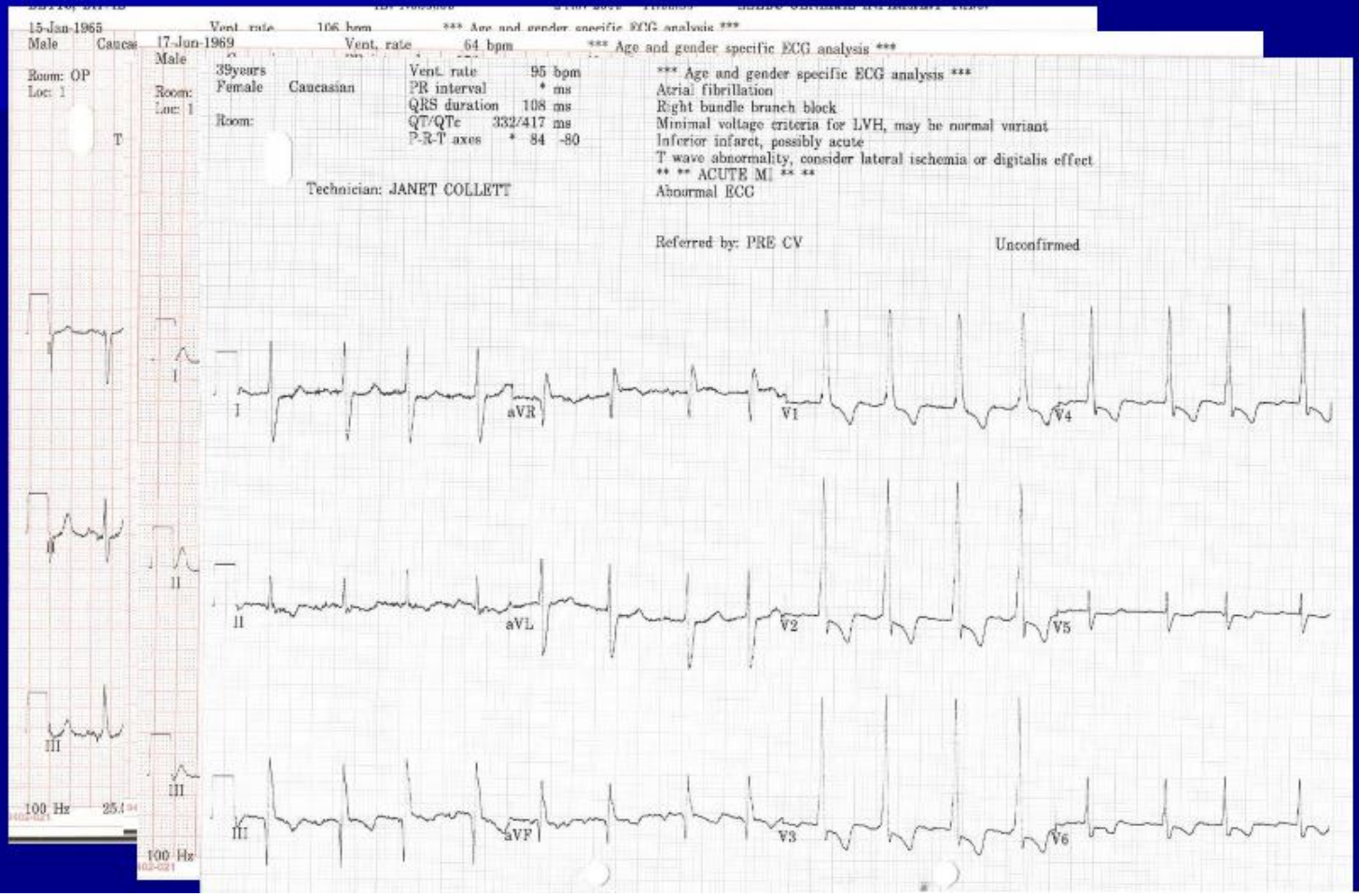
Thrombosed Metal Valves

- High index of suspicion
- Pregnancy
- Small, old valves
- Poor compliance with anti-coagulation (or poor advice!)
- Treatment depends on circumstance
  - Re-anticoagulate
  - Thrombolyse (high risk of stroke in L sided valves)
  - Surgery

Chest pain

- Common in any congenital heart disease patients (? Scar related, ? Psychogenic)
- Most groups of patients are at no increased risk of developing coronary artery disease (except coarctation, arterial switch)
- Assessment as for anyone with chest pain (common sense risk assessment)
- In Eisenmenger patients chest pain may represent RV angina
- Take care in patients with coarctation repairs
  - ? dissection or aneurysm

Beware the abnormal resting ECG!



It may be normal for the patient. All patients encouraged to keep a copy of their own ECG!

Non-Cardiac Emergencies

- Often general surgical, orthopaedic, gynae
- General principles of the management of cyanotic patients (fluids, filters etc)
- Extra care with general anaesthesia - particularly Fontans and eisenmengers
- Care with central venous access - central veins often scarred / occluded (VVs)

https://youtu.be/zQkSiavqbLs

- They have finely balanced haemodynamics
- danger of over-diuresis + vasodilators
- “left heart”(systemic) heart failure uncommon (except systemic RVs)
- More likely to see chronic poor systemic cardiac output and “right heart”(sub-pulmonic) failure
- Right Heart failure more common. Sub acute with anatomic/functional decline
- Acute onset ventricular dysfunction check for:
  - arrhythmic
  - new anatomic substrate
  - infection
  - ischaemia

https://youtu.be/lgXqJnE4hFk

Care of the cyanosed patient

- Avoid iron deficient anaemia
- Avoid dehydration
- Avoid paradoxical emboli/air embolism
- Avoid inappropriate anticoagulation
- At risk of bleeding – haemoptysis in PAH
- Avoid infection
- Be cautious of new medications that modify pulmonary or systemic vascular resistance

- Adaptive mechanism
- Isolated increase in red cells and Hb
- Cyanosis may be missed in a patient with oxygen saturations of 85% who has an Hb of 110g/dL
- In fact their Hb should be around 200g/dL!
- Oxygen delivery to the tissues is maximised when the Hb is appropriately high
- Make sure the Hb in a sick cyanotic patient is appropriately high

Avoid iron deficiency in cyanosed patient

- Cyanosed patients at increased risk of iron deficiency
- Caused by inappropriate phlebotomies (not recommended)
- Iron deficient red blood cells – more likely to clump together
- Associated with increased risk of stroke and death
- Iron deficiency can mimic hyper viscosity syndrome – headache, fatigue, restless legs, blurred vision
- IV Iron preferred method of replacement

Care of the cyanosed patient

- Increased risk of stroke + TIA
- Paradoxical emboli
- Triggered by dehydration
- Triggered by Iron deficiency – strong predictor for ischaemic cerebrovascular events
- Arrhythmias – increased risk
- PPM/ICD – risk of thrombus + stroke

IV Filters

To be used in all patients with a bi-directional or right to left shunt

24hr filter

96hr filter for use with Hickman/PICC lines

24hr Filter for Lipids/Parenteral Nutrition

IV Filters continued..

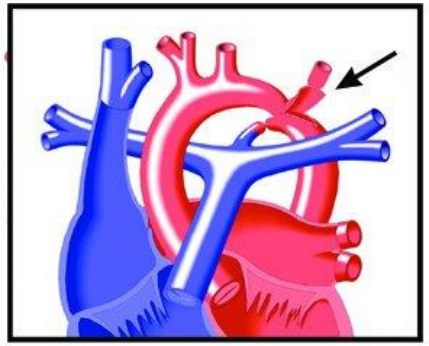
- Contamination can have serious consequences
- Particles cause phlebitis and have serious systemic effects, damaging the lung and organs
- Bacteria can grow rapidly in infusion fluids
- Endotoxins have been shown to penetrate conventional IV filters
  - Only filters that retain endotoxins (0.2micron) can safely be used for more than 24 hours
- Air embolism can be fatal

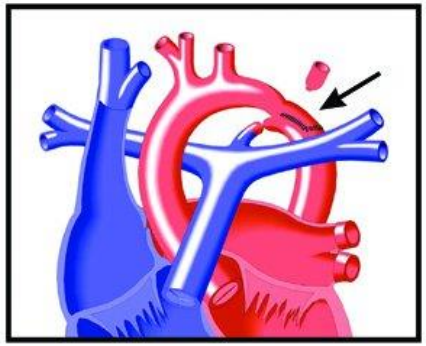
- Cyanotic patients are at particular risk
- Limited cardiovascular reserve to cope with infection
- New headache is always suspicious of cerebral abscess
- IV filters important

Special consideration of cyanosed patients with high haematocrit

- Corrected citrate once haematocrit >0.550
- Need FBC first to calculate haematocrit
- Amount of citrate adjusted according the haematocrit – individual to each patient
- Risk of abnormal results if do not use
- Dangerous for patient

- Be careful – think about which arm to use
- BT Shunts – avoid affected arm
- Coarctation – right arm only
  - Sub-clavian flap repair
  - Left radial pulses weak
  - Left arm B/P artificially low + unreliable
  - Right arm/leg B/P gradients





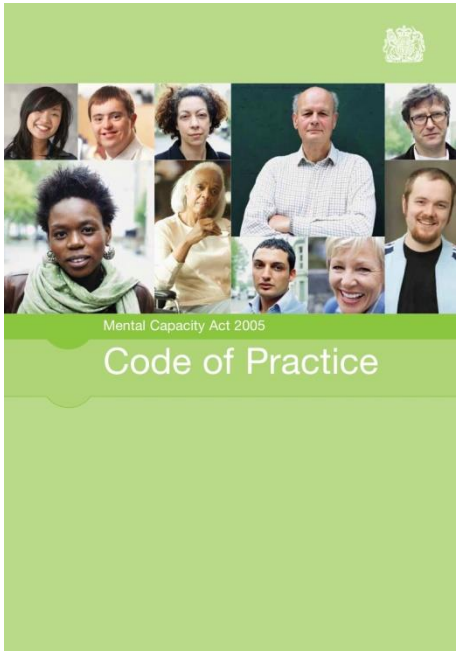
Di-George Syndrome

- Also known as CATCH22, or 22q11 micro-deletion or velofacial cardio syndrome
- Low immunity
- Although low risk – host versus graft disease following blood transfusion
  - Universally fatal!
- Always need irradiated blood and CMV negative
- http://www.maxappeal.org.uk/

The mental capacity Act

Mental Capacity Act 2005

- A significant number of patients will have learning difficulties.
- It is your duty to understand the Mental Capacity Act 2005 and how it applies to each situation
- You are legally required to know the code and to abide by it



https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/497253/Mental-capacity-act-code-of-practice.pdf

Remember!

- ‘Repaired’ does not mean normal – either structurally or physiologically
- You won’t know what all the different operations are and what their importance is
- Common sense and cautious approach is invaluable
- The patient is likely to know much more than you – be prepared to listen

In Summary

- Know your anatomy!
  - 1 or 2 ventricles
  - Systemic RV or LV
  - Pressure and volumes may be critical
- Understand relevance of altered clinical observations
- Know what's normal for your patient
- Special considerations for cyanosed patients and patients with a systemic or single ventricle
- Have a low threshold for seeking expert help
- Know how to contact the ACHD Team!

Contact Details ACHD Team

ACHD Helpline: 0161 276 7959

ACHD on call via switch board:

Before September 2018: 0161 276 1234 and ask for Cardiology middle grade on call

After September 2018: contact details may change but staff will be informed