6pm - On call shift

Trauma SHO pops into theatre

- Booking form for a 3 year old who has a fractured distal radius
- He Is starved
- The registrar saw him but didn’t mention any specifics
- SHO heard mum mention a Glenn shunt
- They would like to proceed with an MUA and possible K-wire as soon as possible.

Alarm Bells

What else do you need to know?

- Glenn Shunt?
- Urgency of Surgery?
- Are we able to do the surgery?

Glenn Shunt

What is it?

- Second stage of palliation surgery for a single ventricle patient heading towards a Fontan circulation.
- Aim to divert systemic venous return via pulmonary vasculature without overloading it.
- Balanced circulation requires complex knowledge of physiology of defect and effects of anaesthetic.
- Requires discussion with Paediatric specialist centre.
- Will likely require transfer for surgery

Paediatric cardiac disease in non-cardiac surgery.

A basic approach

Richard Hayes

Aims

NOT A COMPREHENSIVE GUIDE TO ALL CARDIAC DISEASE

- How to classify and Grade Heart disease
- Who to call for advice
- When to cancel
- Case based discussions

Where to Start?

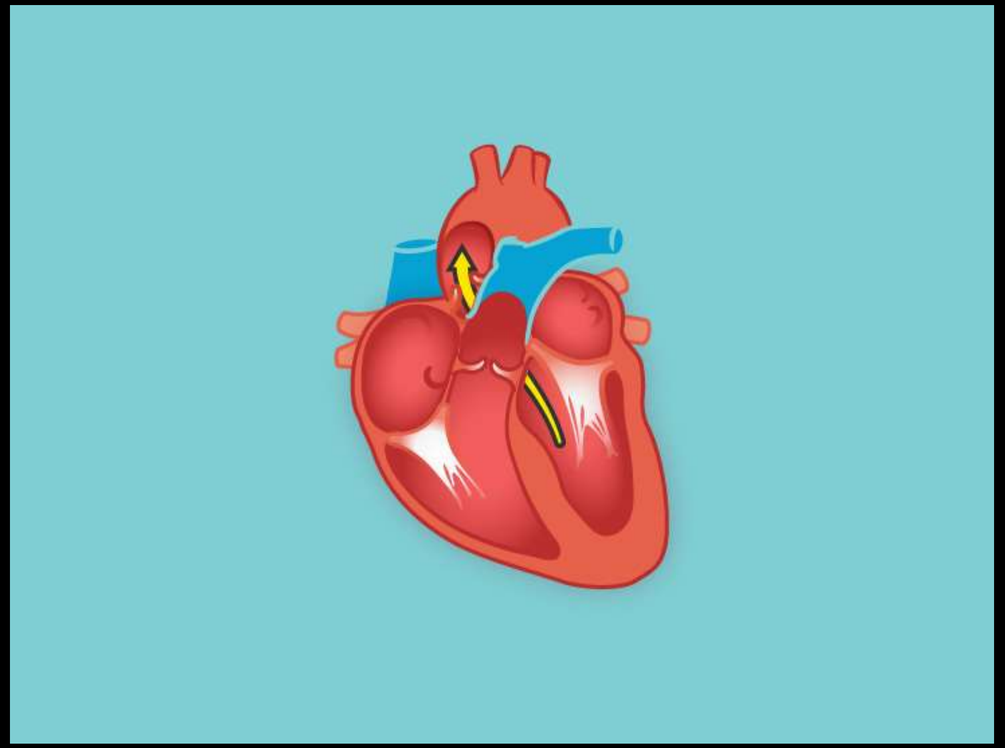
Background figures for CHD.

- 1 in 125 live births
- 90% survival to adulthood
- Present for elective and emergency surgery
- Higher risk of peri-operative cardiac arrest and Higher 30 day mortality
- Variety and complexity of defects and risk require a case by case assessment

Different Types of Circulation

How does blood flow and mix?

- Normal or ‘series’ circulation
- Balanced Circulation
- Single ventricle circulation



The Normal “Series” Circulation

Separate systemic and pulmonary circulations working in series

- Most types of repairs CHD
- Some types of unprepared CHD have this circulation but with holes allowing mixing of blood eg. ASD or VSD
- Blood flows through the hole down a pressure gradient
- Left to right shunts result in increased pulmonary blood flow and potentially decreased systemic blood flow.
- Right to left shunts lead to deoxygenated blood bypassing the pulmonary circulation and therefore decreased pulmonary blood flow and cyanosis.

The Normal “Series” Circulation

Separate systemic and pulmonary circulations working in series

- Amount of shunting depends on pressure gradient
- Changes in PVR and SVR from anaesthesia and administration of oxygen has greatest effect on large unrestricted defects.
- Infants with large unrestricted defects can exhibit “Balanced” Circulation physiology

Parallell or “Balanced” Circulation

Pulmonary and systemic circulations communicate with each other and function as being parallel

- Anatomical abnormalities cause blood flow to systemic and pulmonary circulation to vary depending on the relative resistances in each circuit.
- Blood flow to lungs and body is a balance of PVR and SVR
- Excessive PBF causes Oedema and decreased systemic perfusion which can profoundly affect coronary and splanchnic perfusion.
- Insufficient pulmonary blood flow leads to profound cyanosis

Parallell or “Balanced” Circulation

Pulmonary and systemic circulations communicate with each other and function as being parallel

- A local hospital may see infants with large unrepaired AVSD or VSD.
- Predominantly L-R shunt
- High O2 conc can increase pulmonary blood flow and reduce systemic perfusion.
- High dose induction agent may decrease SVR to the point of shunt flow reversal causing desaturation
- Excessive Pulmonary blood flow leads to risk of pulmonary hypertension.

Parallell or “Balanced” Circulation

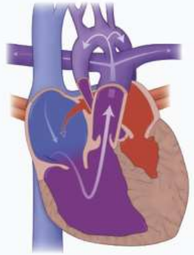
Pulmonary and systemic circulations communicate with each other and function as being parallel

- Other examples
- Modified Blalock-Tausig shunt
- Truncus arteroisus
- And hypoplastic left heart syndrome
- Can be difficult to manage and require discussion with regional paediatric cardiac centre

Single-ventricle Circulation

Not amenable to biventricular repair resulting in series circulation

- Single ventricle pumps blood to systemic circulation.
- Blood flows passively down a pressure gradient from venous system through pulmonary circulation
- There are three stages to formation of a single ventricle circulation
- 1. Formation of a Blallock-Taussig Shunt - small tube connecting arterial to pulmonary circulation to allow a small amount of extra blood into pulmonary circulation.
- 2. Glenn shunt - SVC connected to pulmonary artery
- 3. Fontan procedure - IVC also connected to pulmonary artery - the venous return goes straight to lungs bypassing the heart and the single ventricle pumps blood purely to the systemic circulation.



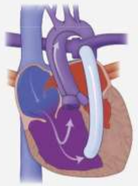
Transplant | **Palliation** | Corrective surgery

STAGE 1

Norwood

- Ligation of PDA
- DKS anastamosis & arch reconstruction
- Atrial septectomy
- Systemic to pulmonary synthetic shunt:
  - a) Sano shunt
  - b) Modified-BT shunt

a)



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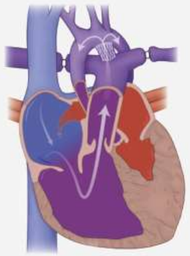


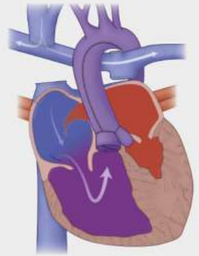
Hybrid

- Percutaneous stenting of ductus arteriosis
- Bilateral PA banding

STAGE 2

Bidirectional glenn

- Takedown systemic to pulmonary shunt
- SVC to RPA anastamosis



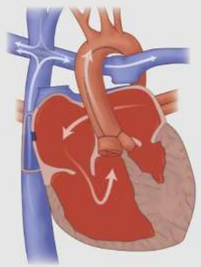
Comprehensive stage 2

- Removal of PDA stent and PDA ligation
- Branch PA debanding
- DKS anastamosis & arch reconstruction
- Atrial septectomy
- SVC to RPA anastamosis

STAGE 3

Fontan/total Cavopulmonary correction

- Intra or extracardiac IVC to RPA anastamosis



Single-ventricle Circulation

Not amenable to biventricular repair resulting in series circulation

- Pressure gradient from pulmonary artery to Left atrium is the sole determinant of pulmonary blood flow.
- Increases in PVR and Intrathoracic pressure can compromise Pulmonary blood flow.
- Spont ventilation causes negative intrathoracic pressure and thus can increase pulmonary blood flow.
- Conversely PPV can give greater control of oxygenation and minute ventilation thus avoiding hypoxia and hypercapnia.

Assessment of Risk

No simple algorithm

- Combination of
  - Age
  - Complexity of disease
  - Physiological Status
  - Type of Surgery
- A physiologically well compensated child with CHD undergoing elective surgery can be low risk
- Poorly compensated patients undergoing urgent or emergency surgery carry high risk

Physiological status

4 major risk factors

- Cardiac Failure
- Pulmonary Hypertension
- Arrythmias
- Cyanosis

Cardiac Failure

Signs differ across ages

- Common - Tachypnoea, tachycardia, sweating and cool peripheries
- In Infancy - poor feeding, failure to thrive and hepatomegaly
    - The heart can be volume overloaded, pressure overloaded or a combination.
    - Severe cardiac failure carries very high Risk.
    - In emergency situation retrieval may be required
    - If very mild and asymptomatic could use gas induction or ketamine with second experienced anaesthetist.

Pulmonary Hypertension

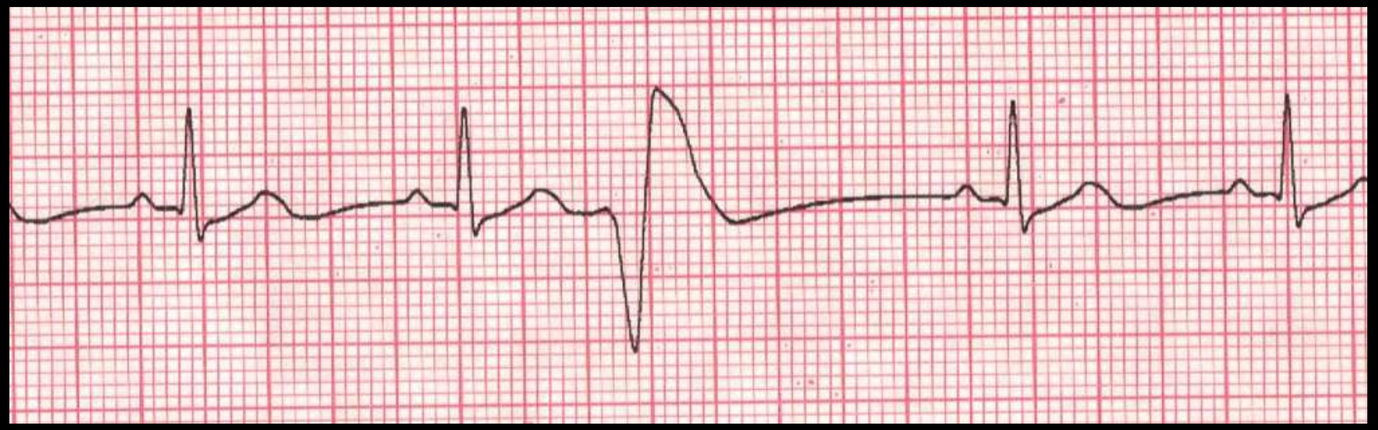
PA Pressure > 25 mmHg at rest and >30mmHg during exercise

- Documented PHT is a clear predictor of periopertive morbidity
- 8 x more likely to suffer a major complication
- Rx with 100% O2, nitric oxide, I.v. prostacyclin, inotropes to support right heart and other measures may be needed.
- Needs transfer to tertiary unit with PICU

Arrythmias

A different story in children

- All children with CHD need ECG pre-op
- RBBB common, unlikely to deteriorate into CHB
- VE’s are a red flag - 30% children with VEs die suddenly - need tertiary centre
- 30% patients with single ventricle will suffer a fatal arrhythmia



Cyanosis

Common feature of unprepared or partially palliated CHD

- Usually concurrent cardiac failure, PHT and arrhythmia - very high risk group
- Paper states that could have minor procedures locally if disease well understood by all staff - likely not feasible.
- Chronic cyanosis leads to polycythaemia and coagulopathy
- Under 5s can get cerebral vein and sinus thrombosis
- Dehydration and iron deficiency anaemia add to risk
- Aspirin should continue if they are on it

Complexity of disease

Complex disease increases risk

- Single ventricle physiology
- Balanced circulation physiology
- Cardiomyopathy
- Aortic stenosis
- Long term sequelae - cardiac failure, PHT, arrhythmia and cyanosis

Type of surgery

More invasive - more risk

- Mortality in major surgery 16% minor surgery 3%
- Major = intraperitoneal, intrathoracic or vascular reconstructive surgery, any surgery where blood transfusion may be required.
- Surgery with prolonged hospital stay also carries high risk

Risk Management

High Risk	Intermediate Risk	Low Risk
Physiologically poorly compensated and/or presence of major complications Cardiac Failure Pulmonary Hypertension Arrythmias Cyanosis	Physiologically normal or well compensated	Physiologically normal or well compensated
Complex lesions (Single ventricle or balanced circulation physiology, cardiomyopathy, aortic stenosis)	Simple Lesions	Simple lesions
Major Surgery (Intraperitoneal, intrathoracic, anticipated major blood loss)	Major Surgery (Intraperitoneal, intrathoracic, anticipated major blood loss)	Minor or body surface surgery
Under 2 years old Emergency Surgery Preoperative hospital stay >10 days ASA IV or V	Under 2 years old Emergency Surgery Preoperative hospital stay >10 days ASA IV or V	Over 2 years old Elective surgery Pre-operative hospital stay <10 days ASA= I - III

Risk Management

Elective procedures

- High risk - transfer to specialist centre
- Intermediate risk - discuss with specialist centre and consider transfer
- Low risk - could be managed at local hospital

Risk Management

Emergency procedures

- High risk and Intermediate risk - discuss with specialist centre and consider transfer
- Low risk - could be managed at local hospital with input from specialist team.

Case Studies

Time for some discussion!!

9 Year Old

RIF Pain and Vomiting

- VSD repair as Infant
- Sent home from mainstream school after games lesson where he was playing football and then developed abdominal pain and vomited, he has had fevers since.
- Surgeon has diagnosed appendicitis and would like to operate that evening
- Is it safe to proceed?
- What do we need to know? What sources of information are there?

3 year old

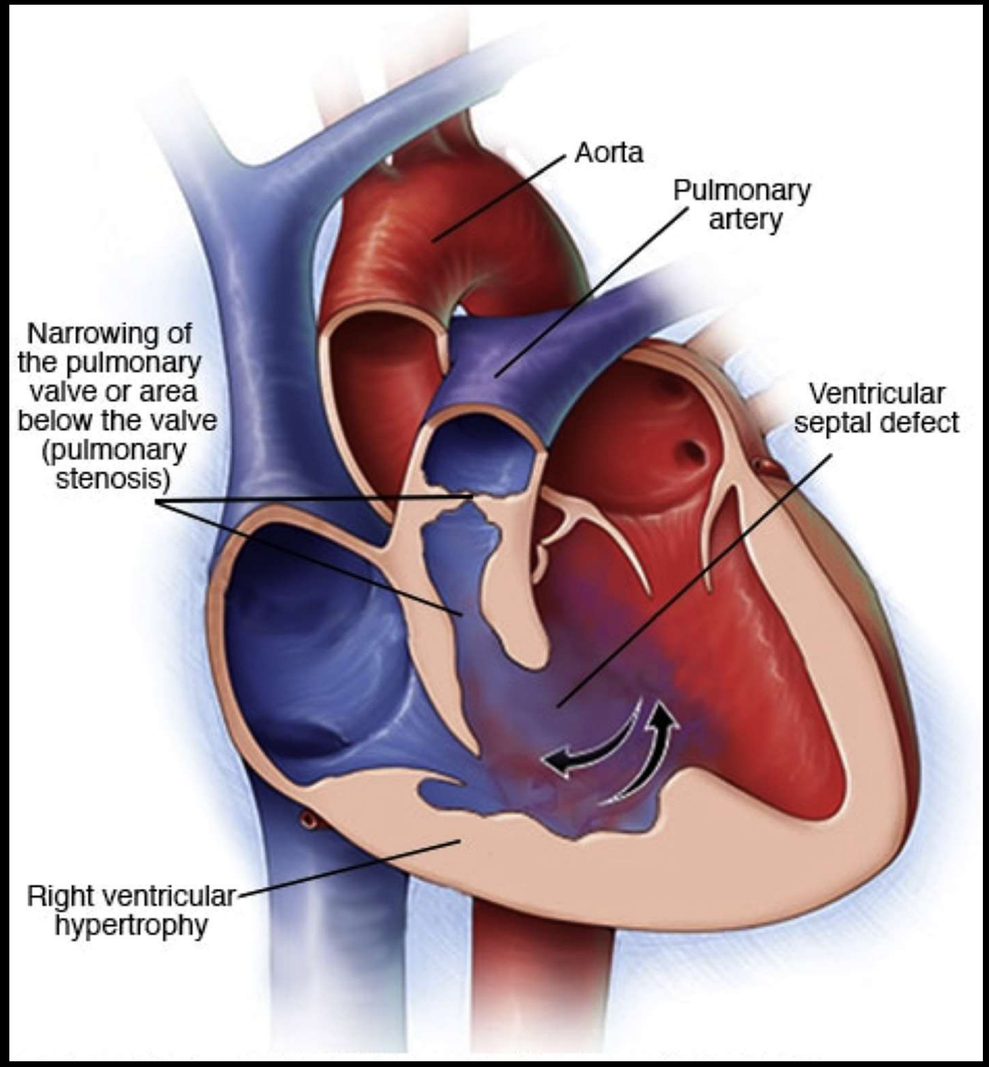
Trisomy 21

- Scalp laceration after falling into a coffee table - full thickness requiring washout and suturing.
- Mum mentions Tetralogy of fallout
- What is this?
- Where can you get information?
- What features may you see?

Tetralogy of Fallot

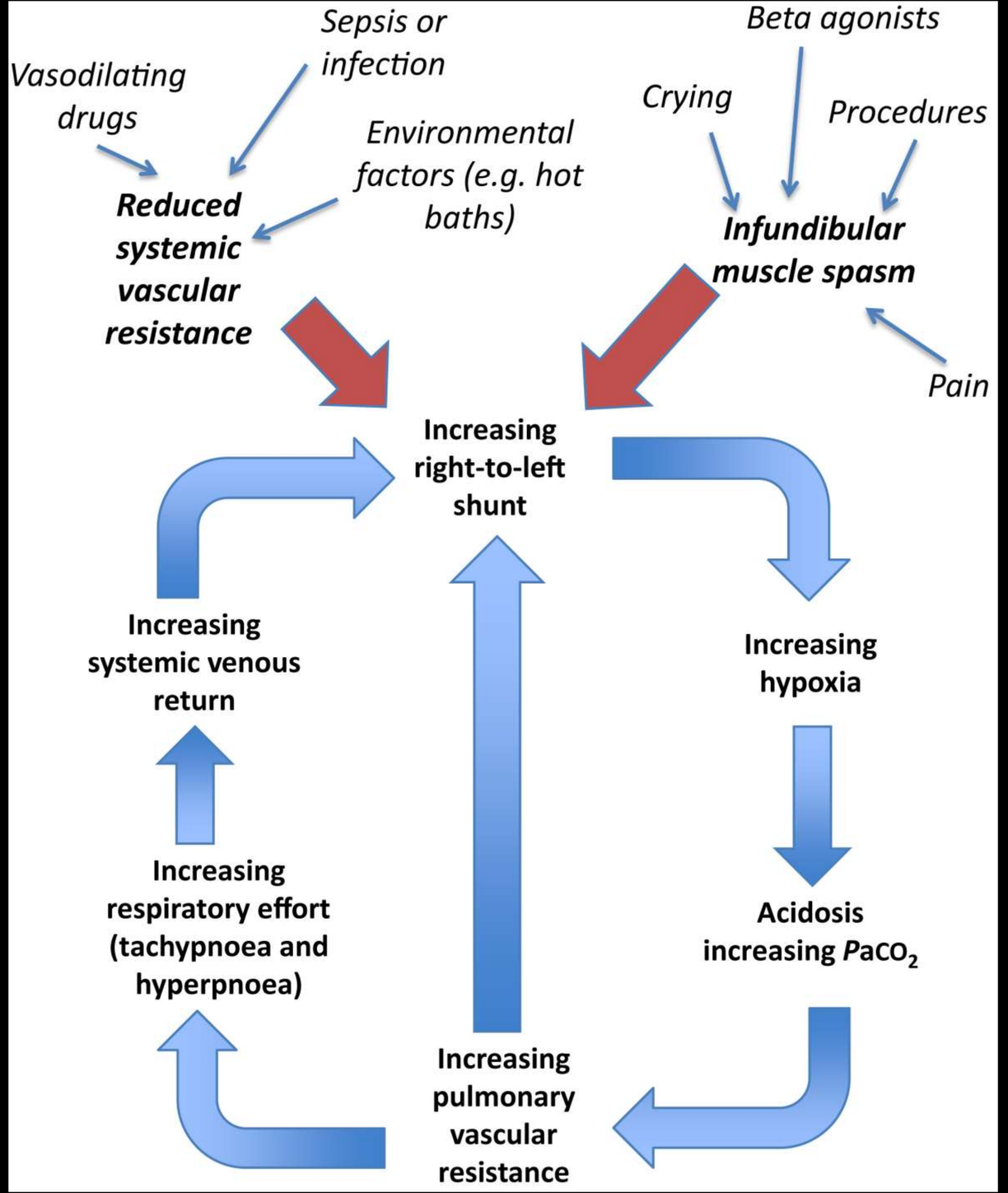
Common cyanotic congenital heart malformations

- Cardinal features are VSD, RVOTO, Overriding Aorta and RV hypertrophy
- Anatomy allows blood mixing between pulmonary and systemic circulations - R-L shunt - deoxygenated blood mixes with oxygenated blood.
- If RVOTO increases due to muscle spasm in times of stress R-L shunt flow increases in a cyanotic spell
- Drops in SVR also increase R-L shunt
- Older Children Squat in knee to chest position to Increase SVR



Increasing hypoxia
Increased degree of RVOTO
Increasing amount of aortic override
Increasing right-to-left shunt

‘Pink’ Tetralogy of Fallot	‘Blue’ Tetralogy of Fallot	‘Profound cyanosis’ Tetralogy of Fallot
No or minimal RVOTO with little aortic override. Behaves like a VSD with left-to-right shunt	Classic ToF has a degree of RVOTO and aortic override	Severe or complete RVOTO


Sepsis or infection
Beta agonists
Vasodilating drugs
Crying
Procedures
Environmental factors (e.g. hot baths)
Reduced systemic vascular resistance
Infundibular muscle spasm
Pain
Increasing right-to-left shunt
Increasing systemic venous return
Increasing hypoxia
Increasing respiratory effort (tachypnoea and hyperpnoea)
Acidosis increasing PaCO2
Increasing pulmonary vascular resistance

2 Year old

Incarcerated inguinal hernia

- Pale and sweaty, tachypnoeic, hepatomegaly on abdominal examination, not mobilising yet.
- Has neonatal marfans syndrome
- Multiple hospital admissions with Chest infections
- Mum brings a lever arch file with notes from multiple different hospital admissions
- Came to nearest hospital as child distressed and had breathing difficulty
- What do you do?

Summary

- The CHD child presenting for non cardiac surgery poses many challenges
- Information is key
- Parents usually well informed
- Anaesthesia poses great risk as disrupts the balance of systemic and pulmonary vascular resistance along with myocardial contractility
- Assessment of risk will guide where surgery should happen
- If in doubt discuss with hospital paediatric lead or local paediatric centre

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