



# Palliative Care and Paediatric Cardiac Patients

5<sup>th</sup> Annual APPM Paediatric Palliative Care Study Day

Dr Caroline Jones

Consultant Fetal and Paediatric Cardiologist Alder Hey Children's Hospital, Liverpool

Alder Hey Children's   
NHS Foundation Trust



# Background

- Congenital Heart Disease (CHD) remains the commonest congenital abnormality and leading cause of birth defect-related mortality
- Prenatal Diagnosis is increasing nationwide
  - A significant proportion with unreparable CHD will opt for TOP
- Outcomes following CHD surgery continue to improve for the majority, more infants are undergoing primary biventricular repair
- Small subset with complex disease remain difficult to treat, 'palliative surgery' remains only option
- In some with difficult morphology, small size, prematurity or significant co-morbidity 'palliation' may not be appropriate or in the child's 'best interests'



# Background

- Historically cardiology had few links with palliative care teams though this is changing
- Cardiologists have been reluctant to refer patients onto PC services
- In older children referral point can be difficult as many may live for years with ongoing symptoms prior to death
- Death in children with CHD is then often sudden and unpredictable
- Many children, teenagers and young adults are in the community with untreated CHD without access to PC or associated services
- In the next decades there will be increasing needs for PC for young adults with CHD



# Decision making in paediatric cardiac care

- Refusing surgical treatment or intervention is always difficult
- A quantitative (scientific) definition Schneidermann (1994) asserts that if an intervention does not 'work' in more than 1% of attempts, it should be considered futile
- Looser ethical definition : a treatment is futile if its intent or effect is to prolong dying without much benefit to the patient
- Who decides what is beneficial? Sanctity of life. We all act in the 'best interest of our patients' don't we?
- Parental views are evolving and the many want every treatment possible
- Importance of MDT decision making and seeking a 2<sup>nd</sup> opinion on behalf of the parents where appropriate



# Managing futility in critically ill patients with cardiac disease.

Price S. Haxby E. Nat Rev Cardiol. 2013 Dec;10(12):723-31

Cardiologists & intensivists often regard death as failure, continuing to pursue active treatment while potentially denying patients access to alternatives such as symptom control and end-of-life care.

Patient autonomy is central to the delivery of high-quality care, although many cardiologists and intensivists do provide thoughtful and patient-centred care, the pressure to intervene can lead to physician-centric care, focused around the needs and wishes of medical staff to the detriment of patients, families, health-care workers, and society as a whole.



# Patients where conventional treatment may not be appropriate or successful

## **Patient Factors**

- Prematurity
- Small size
- Significant chromosomal or genetic abnormality
- 'Significant' co morbidity
  - Reduce CHD surgery survival
  - Poor QoL expected

## **CHD/ cardiac factors**

- Single ventricle patients
  - Abnormal pulmonary veins
  - Severe AV valve regurgitation
  - Impaired vent function
- Pulmonary hypertension untreatable with medical therapies
- Cardiac dysfunction assoc with generalised myopathy
- Patients not suitable for cardiac transplant

# Even with Advancements, Some Things Just Can't be Fixed

Crit Care Med 1993;21:1798-802.

Cases where care was discontinued in PICU

- 19% Congenital heart disease
- 18% Trauma
- 14% Pneumonia/ sepsis
- 14% Anoxia/drown
- 9% SIDS
- 26% Other

In contrast to other specialities, surgical treatment may not be offered rather than medical treatment failing to work

## The Telegraph

Minutes-old baby is UK's youngest heart op patient

Chanel Murrish undergoes open-heart surgery within minutes of being born after a scan convinces her parents to refuse to terminate the pregnancy



# Understanding the physiology will help in treating the symptoms

- Acyanotic
  - pink and breathless
- Cyanotic
  - blue and exercise intolerant
  - High Hb susceptible to thrombosis, stroke
  - Pulmonary hypertensive
- Haemodynamic characteristics
  - Increased pulmonary blood flow
  - Decreased pulmonary blood flow
  - Obstruction of blood flow out of the heart
  - Mixed blood flow
- Breathlessness/  
Dyspnoea
  - Cyanosis
  - Feeding
  - Exercise capacity
- Cough
- Haemoptysis
- Oedema, ascites, pleural effusions

# Paediatric Indicators of Cardiac Dysfunction

- Poor feeding
- Tachypnoea/tachycardia
- Failure to thrive/poor weight gain/activity intolerance
- Developmental delay
- Prenatal history
- Family history of cardiac disease



# Heart Failure treatment options

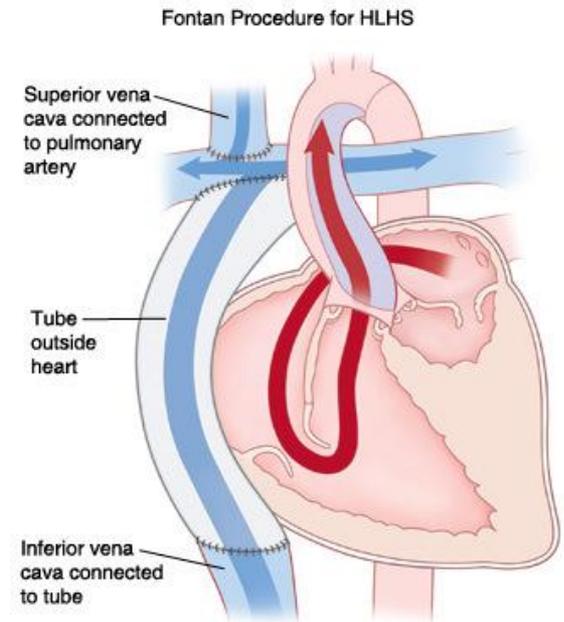
- Diuretics (Symptom Control)
- Vasodilators (Symptom Control)
- Beta blockers (Improve Outcomes)
- ACE therapy (Improve Outcomes)
- Spironolactone (Improve Outcomes)
- Digoxin
- In the palliative care setting rationalisation of medications to those that improve symptoms may be appropriate



# Fontan circulation: success or failure?

Mondesert et al. 2013, Canadian Journal of Cardiology

- Not curative
- 'Health status of children and adolescents is poor'
- 90% survive or do not need a heart transplant 10 years post-Fontan, whereas these numbers drop to 83% 20 years and 70% 25 years post-Fontan
- Patients die of heart failure, stroke or SCD
- Progressive cyanosis, exercise intolerance and development of cardiac arrhythmia
- May have symptoms and signs of 'functional' SVC obstruction
- Creation of a new disease entity
  - Protein losing enteropathy
  - Liver dysfunction/failure
  - Plastic bronchitis
- Reality is transplant only available to a minority of patients



## Forty Years of The Fontan Operation: A Failed Strategy

# Breathlessness or Dyspnoea

- May be multifactorial
  - CHD, anaemia, pleural effusion, airway obstruction
- Susceptible to intercurrent infections
- Anxiety provoking and may confound the problem increasing cardiovascular demand
  - Low dose midazolam, chloral hydrate, opiate
- Orthopnoea – sleeping sitting up may helpful
- Low flow oxygen may be beneficial in some cases
  - If there is mechanical obstruction to pulmonary blood flow this is unlikely to help

# Cough

- Treat underlying causes
  - Heart failure – diuretics
  - Pulmonary hypertension
  - Plastic bronchitis
    - Chest Physiotherapy
    - Bronchodilators
    - N-Acetylcysteine, DNase
    - In paediatric patients with plastic bronchitis secondary to Fontan physiology, 45% of the 18 reported cases have died from asphyxia secondary to airway obstruction
- Humidified air or oxygen, neb saline
- Linctus, codeine



# Haemoptysis

- This can be particularly distressing for the family and possibility of this should be discussed
- May be seen in pulmonary hypertensive patients or group with pulmonary atresia/ MAPCAs
  - Tranexamic acid
  - Nebulised adrenaline
- Massive pulmonary haemorrhage – unusual
  - Buccal or intranasal midazolam or morphine repeated every 10 minutes until the child is settled
  - May cause rapid death in children



# Summary

- Complex forms of CHD remain unreparable though results of 'surgical palliation' are improving
- Symptoms in end-stage CHD may be challenging to treat
- Continued strengthening of links between palliative care and paediatric cardiology teams will improve patient care in this group of patients
- Patients and families have a huge amount to gain from your specialist skills, support and knowledge
- Likely that we should consider referring patients earlier
- Expect more referrals!

