Palliative Care and Paediatric Cardiac Patients

5th Annual APPM Paediatric Palliative Care Study Day

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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 unrepairable 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 2nd opinion on behalf of the parents where appropriate.
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


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
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
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!