PICU management Heart Failure

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Objectives

• Identifying common causes of heart failure in children

• Diagnoses of cardiac lesions associated with heart failure

• ICU management plan
What are the common causes of heart failure in children?

- **Congenital Cardiac Diseases**
- **Acquired cardiac diseases**
  - Cardiomyopathy
  - Myocarditis (usually with CHF)
  - Pericarditis
  - Arrhythmias
  - Kawasaki
  - Toxic (adriamycin)
- **In association with systemic diseases or syndrome**
Approximately 1% of infants have CHD.

Common symptoms at presentation
1) Heart Failure
2) Cyanosis
3) Heart murmur
4) Apnea
5) Cardiogenic shock
Other causes of failing dilated heart in children

<table>
<thead>
<tr>
<th>&lt; 1 YEAR OLD</th>
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<td>18. Nemaline Myopathy</td>
<td>18. Peripartum Cardiomyopathy</td>
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<td>19. Minicore-Multicore Myopathy</td>
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<td>20. Myotubular Myopathy</td>
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Case Scenario

- 2 weeks old infant brought in by parents with difficulty breathing
- HR 180, BP 50/P, RR 80, T 37.5
- Poor feeding and tachypnea since birth
- Pale with mottled skin
- Delayed capillary refill
- Poor perfusion
Differential Diagnosis of Infant Shock

- **infection** (septic shock/ meningitis)
  - bacterial: GBS, E. coli, S. aureus
  - virus: enteroviruses, H. simplex
- **metabolic**: amino/organic acidopathies, urea cycle defect
- ‘**hypoxic shock**’: eg. RSV, C.N.S disease
- **heart disease**: congenital or acquired
A 10 days old infant was referred to our ICU because of respiratory distress progressed rapidly to shock state.
LV Outflow Obstruction

- Coarctation of the aorta
- Hypoplastic left heart syndrome
- Aortic stenosis (presents later)

- “Any neonate in shock that does not respond to fluids or pressors has LV outflow obstruction until proven otherwise”

- Complete obstruction incompatible with life unless there is shunting
Coarctation of the Aorta

{Most common cause of heart failure in the first month of life}

Most often distal to L subclavian
Management of left sided obstructive lesions

- Stabilization, PGE2
- ASAP surgical repair
Case (9): A 2 months old boy presented to emergency room in severe respiratory distress. He required intubation and mechanical ventilation. After intubation CXR was obtained.
VSD

{Left to Right Shunt}

• Most common congenital lesion
• Large VSD’s may be silent and become symptomatic in first few weeks as pulmonary resistance ↓
  • SOB and diaphoresis with feeds
  • Poor weight gain

• Systolic murmur
• CXR demonstrates CHF
PDA
Increase pulmonary blood flow from left to right shunt
ICU management of intractable heart failure

- Diruetics, afterload reduction, positive pressure ventilation
- Patients with significant left to right shunt may present in:
  - Intractable heart failure
  - Required prolong intubation
  - Severe respiratory infection that can not be cleared
  - Persistent patchy lung infiltrate

All the above mentioned complication may not improve until the L-R shunt is eliminated by surgical repair
Correlation between SD of body weight from the mean and duration of ventilation hours

$r = 0.097$
$n = 79$
Correlation between SD of body weight from the mean and duration of inotrops infusion (hours)

\[ r = 0.025 \]
\[ n = 79 \]
Acquired Heart Disease

- Acquired
  - Cardiomyopathy
  - Myocarditis (usually with CHF)
  - pericarditis
  - Dysrhythmias
  - Kawasaki
CHF - Etiology

• presents immediately at birth
  – anemia, acidosis, hypoxia, hypoglycemia, hypocalcemia, sepsis

• presents at 1 week (congenital)
  – PDA in premature infants

• presents in first month (congenital)
  – HPLV, aortic stenosis, coarctation, VSD presents later

• presents later (acquired)
  – myocarditis, cardiomyopathy (dilated or hypertrophic), SVT, severe anemia, rheumatic fever
Case presentation

- 7 month old with irritability, poor feeding (fatigues), poor weight gain, fast breathing
- no fever or runny nose
- physical exam: HR 160, RR 60, BP 80/40 with minimal respiratory distress, gallop rhythm, rales
- Hepatomegaly
Dilated Cardiomyopathy
Myocarditis

- leading cause of dilated cardiomyopathy and one of the most common causes of acute CHF in children
- etiology: idiopathic, viral, bacterial, parasitic
- hallmark is CHF
- failure to respond to bronchodilators in wheezing child
- treatment includes inotropes, afterload reduction, diuretics.
- IVIG, steroid, immuno-suppresion are controversial
Special attention!!

- Intubation can be nightmare with significant incidence of cardiac arrest during induction.
- CPAP alone, is possible alternative choice if tolerated with caution.
- Negative pressure ventilation is reported to be useful
- Positive pressure ventilation and PEEP help by:
  - Minimizing venous return
  - Decrease pulmonary edema and alveolar capillary leak
  - Decrease WOB and metabolic demands
  - Decrease the left ventricular afterload
Invasive Mechanical Support of Heart

- ECMO
- Ventricular assist device
  - Refractory Cardiac output state
  - Bridge to heart transplant
  - After Cardio-pulmonary resuscitation
  - In myocarditis refractory to medical management
Pericarditis

- sharp stabbing precordial pain
- worse with supine and better leaning forward
- no sensory innervation of the pericardium
  - pain referred from diaphragmatic and pleural irritation
Case presentation

- 7 year old female, 5 days prior to admission.
- Fever, sore throat, non-specific abdominal pain.

- Initial exam revealed stable vital signs, HR 130, RR 25, BP 120/70, Temp 38, SAT 95%, bilateral crepitation, and NON-specific generalized abdominal.

- A CXR in ER showed cardiomegaly and bilateral lung infiltrate V.S congestion.
Etiology

- **infectious**
  - viral
  - bacterial
  - TB
  - fungal
  - parasitic
- **Connective tissue**
  - RA
  - Rheumatic fever
  - SLE
- **Metabolic / Endocrine**
  - uremia
  - hypothyroid
- **Hematology / Oncology**
  - bleeding diathesis
  - malignancy
- **Trauma**
- **Iatrogenic**
What is the immediate priority in this life threatening cardiac emergency?

Decompressing the heart: Pericardiocentesis, followed by broad spectrum antibiotics administration.
Pericarditis

- Usually a benign course
- Virulent bacteria (H. flu, E. coli) can cause constrictive pericarditis and subsequent tamponade – may need urgent pericardiocentesis
- Uncomplicated pericarditis usually responds to rest and anti-inflammatories
Infective Pericarditis although is rare disease but not absent.


**Five children with purulent pericarditis and review of the literature**

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Pediatric arrhythmia
Pediatric dysrhythmias

• Fast or slow rhythm
• Often they are caused by underlying CHD
• After cardiac surgery
• In previously healthy child consider
  - Drug toxicity
  - Electrolytes imbalance \{K, Mg, Ca\}. 
9 Year old with frequent episode of palpitation, ECHO showed thrombus in R-atrium
Management

- BLS
- O2, {12 leads ECG if possible}
- Unstable: Cardioversion {0.5-1 J/kg}
- Stable: (normal BP, Normal perfusion)
  - Vagal stimulation, apply ice bag on forehead
  - IV/IO. Adenosin: 0.1 mg /KG {MAX 6 mg}
Ventricular Tachycardia

QRS > 0.08 Seconds
Management of tachyarrhythmia

- BLS
- ABC
- O2
- Attach monitor / defibrillator
- Assess ECG
- No pulse: defibrillation 2 J/Kg
- With Pulse: Cardioversion 0.5-1 J/Kg
- Consider medication: Amiodarone 5 mg/kg
Bradyarrhythmias

• Etiology
  – hypoxia, acidosis, hypoglycemia
  – excess vagal stimulation (ex. intubation)
  – Cold stress, deep sedation
  – Heart block: congenital or after cardiac surgery

• Treatment
  – Epinephrine
  – Atropine if known vagally mediated or heart block
  – Transcutaneous pacing and pacemaker
Congenital Bradyarrhythmias

• complete AV block
  – autoimmune injury to fetal conduction system secondary to maternal autoimmune disease
  – atropine, isoproterenol, epinephrine may be tried temporarily prior to pacing
Kawasaki Disease

- vasculitic process
- diagnostic criteria (5/6)
  - fever > 5 days
  - oral mucosa changes
  - non-purulent conjunctivitis
  - lymphadenopathy
  - rash
  - peripheral edema
- non-specific-hydrops, uveitis, thrombocytosis
Periungual **desquamation** on day 12 of illness in a three-year-old child with Kawasaki disease.

**Rash** on day 4 of illness in a seven-month-old boy with Kawasaki disease.

Edema and erythema on day 6 of illness in a one and one-half-year-old girl with Kawasaki disease.

Lip edema and erythema on day 6 of illness in a two-year-old boy with Kawasaki disease.
Kawasaki Disease

- Pink eye
- Oral mucosal change
- Enlarged lymph nodes
- Patchy rash
- Peeling skin

Coronary artery aneurysms (in 20% of cases)

Inflammation within the heart muscle (in 20% of cases)
Kawasaki--Coronary Aneurysms

Right System

Left System
Kawasaki Disease

Treatment-- to prevent cardiac sequelae

• **Intra Venous Gamma Globulin**

• **Aspirin--antiplatelet dose**
  – persistent coronary dilatation, small aneurisms
Differential diagnosis of Cardiomyopathy

- Coarctation
- ALCAPA
- Missed Kawasaki
Angiogram of Patient with ALCAPA
ALCAPA

EKG Showing Ischemic Pattern

[ECG images showing ischemic pattern]
Cardiovascular - Basic

\[ BP \propto SVR \times CO \]

\[ HR \times SV \]

- Preload
- Contractility
- Afterload
Afterload - Anything Else?

• Two ‘newer’ drugs
• Fenoldopam
• Milrinone
Milrinone

- A phosphodiesterase inhibitor
- Inhibits breakdown of cAMP

Adrenergic Receptors

- α - peripheral vasculature
  Stimulation → vasoconstriction

- β1 - myocardium
  Stimulation - inotropy and chronotropy
  Mechanism: activates adenylate cyclase → cAMP → Ca^2+ entry into the cell → inotropy → chronotropy

- β2 - Lungs and peripheral vasculature
  Stimulation - smooth muscle relaxation, vasodilation and bronchodilation
Milrinone

- As mentioned before a phosphodiesterase inhibitor
- In addition to effects on inotropy and chronotropy
- Potentiates a cAMP dependent vasodilation
  - Not related to adrenergic receptors
- Dose range: 0.3-1 mcg/kg/min
- Use: patients who need both inotropy and afterload reduction
Fenoldopam

- Modification of Dopamine with a phenol group
- Dopamine 1 receptor agonist
- Vasodilation - used in adult hypertension management
- Improved renal blood flow
Nesiritide (B-type natriuretic peptide) in the Treatment of Pediatric Heart Failure

**Fig. 1** Urine output (mean ± SD) before and after initiation of nesiritide infusion ($p = 0.01$). Urine output increased significantly in the first 4 days of nesiritide therapy.
Sudden Cardiac Death
one third of all sudden deaths in children

- Etiology
  - myocarditis
  - cardiomyopathy (hypertrophic)
  - cyanotic and noncyanotic congenital heart disease
  - valvular heart disease
  - congenital complete heart block
  - WPW
  - long QT syndrome
  - Marfan syndrome
  - coronary artery disease
  - anomalous coronary arteries
Principal of PICU heart failure management

Ventricular dysfunction limits a patient's ability to perform the routine activities of daily living...

Diuretics, ACE Inhibitors
Reduce the number of sacks on the wagon

Digitalis Compounds
Like the carrot placed in front of the donkey
Thanks
D-TGA

- 5% OF CHD.
- Most common cardiac cause of cyanosis in newborn.
- VSD is present in 30% of cases
- Newborn with IVS usually have severe cyanosis
TGA (radiological studies)

- Egg-shaped heart
- Narrow waist or pedicle
- Increased pulmonary blood flow
- Cardiomegaly
• PGE1 is life saving

• Balloon atrial septostomy is also life-saving procedure.

• After BAS, Children frequently can be extubated and PGE1 can be stopped
Tricuspid atresia
Obstructed TAPVD
TAPVD, Supra-diaphragmatic

View of abnormal connection of pulmonary veins behind the heart

Oxygenated blood from the right lung

Oxygenated blood from the left lung

RA, LA, PA, RV, LV

TAPVD
TAPVD, Infra-diaphragmatic

Total Anomalous Pulmonary Venous Return

Infradiaphragmatic

AO, PA, LA, RV, LV, RA, PV

Normal
TAPVR
Truncus Arteriosus
Pediatric Cardiac Emergencies are not very common: it can be easily missed or mismanaged:

Possible scenario:

- 1 month old with irritability, poor feeding (fatigues), failure to thrive, fast breathing
- no fever or runny nose
- physical exam: HR 160, RR 60 with minimal respiratory distress, gallop rhythm, crepitation and rales
Why do Congenital Heart Diseases present as emergencies?

- Decrease PVR
- Closure of ASD & PDA
- Establishing pulmonary and systemic circulation
What are the CHD that can present as emergency and why?

**Cyanotic and non-cyanotic heart diseases:**

- Closure of the Ductus in PDA dependent lesions
- Heart failure
- Low cardiac output
- Cyanosis
Cyanotic heart diseases
Cyanotic Heart diseases
When can we recognize cyanosis?

Cyanosis is the result of increased unoxygenated hemoglobin. Usually noticeable if > 20% or more than 5 gram of reduced hemoglobin in capillary bed.
Cyanosis

- Classified as central or peripheral
- Central cyanosis (always abnormal)
  - mucous membranes, trunk, extremities
  - classified as cardiac (R to L shunt) or pulmonary
- Peripheral cyanosis (acrocyanosis)
  - no involvement of mucous membranes
  - involves hands, feet, circumoral area
  - common in neonates from vasomotor instability
  - CHF, PVD, shock, cold extremities
Cyanotic Heart diseases

1- Transposition of Great Arteries
2- Tetralogy of Fallot (↓PBF)
3- Tricuspid Atresia (↓PBF)
4- Total Anomalous Pulmonary Venous Drainage.
5- Truncus Arteriosus
A 4 months old infant with cyanotic heart disease since birth, presented to ER with deep cyanosis, desaturation despite 100% FiO2. The infant is obtunded and limb
• Transient **infundibular spasm** causes increase in right to left shunt and decrease in pulmonary blood flow.

• clinically: desaturation and hypoxia, increase cyanosis, irritability, crying, loss of consciousness

• hypoperfusion and lactic acidosis, syncope, seizure, death.
**Tet spell**

**Pathophysiology:** Hypoxia, dehydration, acidosis, sepsis, agitation............
Management of Tet spell

- Soothing
- oxygen support.
- MSO$_4$.
- NaHCO$_3$
- Maneuver to increase SVR and decrease the shunt: knee-chest position, phenylephrine.
- B-blocker (Propranolol, Esmolol).
- Hydration and correction of acidosis.
- Maneuver to decrease O2 consumption, sedation, relaxation
- Mechanical ventilation, analgesia, sedation
- Surgery
Case Scenario

• 14 year old male collapses at school while in class
• non-responsive for one minute
• feels fine in the ER department
• Approach?
Figure 3. Electrocardiogram demonstrating long QT syndrome. Markedly prolonged QT interval calculated with the Bazett formula: $\text{QTc} = \text{QT} / (\text{square root of the preceding RR interval})$; $\text{QTc} = 0.36/(\text{square root of 0.44})$; QTc = 545 ms. Courtesy of Nicholas Tsarouhas, MD, The Children's Hospital of Philadelphia.
Left ventricular assist device (LVAD)
Biventricular assist device (BiVAD)
Prolong QT Syndrome
Syncope

- 20-50% of adolescents experience at least one episode of syncope
  - most cases benign
- Pathophysiology
  - vascular
  - orthostatic, hypovolemia
  - neurally mediated
  - hypoxia: PE, CNS depression from over dose
  - cardiac
Cardiac Syncope

- Dysrhythmias
  - tachy
  - brady
- Outflow obstruction
- Myocardial Dysfunction
- cardiac syncope often precedes future sudden cardiac death

Figure 2. Electrocardiogram of Wolff-Parkinson-White. Note the delta waves.
SVT before and after treatment

A child presented to ER after episode of chest pain and fainting at home.

notice the Delta shape of QRS in WPW Syndrome
SVT

- HR > 200 per minute (60% > 230)
- Narrow QRS complex < 0.08 second
- Fixed rate \{dose not fluctuate\}
- Difficult to identify P wave
- No beat to beat changes
- Beginning and termination are abrupt