CARDIAC EMERGENCIES

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Consultant Pediatric Cardiologist
Latifa Hospital, DHA
Cardiac emergencies are among the most stressful ED presentations.

Cardiac Problem in infancy & childhood are not rare, often are complex.

Cardiac disease in infancy & childhood can be congenital or acquired.
The most clinically demanding pediatric cardiac emergencies presentation

1. Congestive heart failure
2. Myocardial disease
3. Neonatal cardiac emergencies with obstructed systemic or pulmonary blood flow
4. Hypoxemic attack
5. Cardiac arrhythmias
6. Acute postoperative pulmonary hypertension (pulmonary Hypertensive Crisis)
Heart Failure

A syndrome in which the heart cannot maintain level of tissue perfusion adequate to meet metabolic needs.
CHF can be divided into 4 types based on the underlying pathophysiology:

1. Pulmonary overflow CHF
2. CHF due to Impaired myocardium
3. CHF due to obstructive lesion
4. Tachyarrhythmia induced CHF
Pulmonary overflow CHF

- The most common cause of CHF
  - Excessive volume loads
  - Left to right shunts e.g. VSD, AVSD, arteriovenous malformations

- Precipitating factors exist that lead to decompensation
  - Most common factor is infection, respiratory tract infections

- Mortality rate ~ 40% in infant with CHF with RSV
Clinical Manifestation of acute CHF

1. Respiratory symptoms -- wheezing
2. Tachycardia
3. Abnormalities of cardiac auscultation----Gallop
4. Hepatomegaly
5. Cardiac enlargement
Management

1. Diuretic therapy
2. Oxygen therapy for cases with desaturation
3. Control of infection
4. Digoxin
5. After load reduction
## Management of acute CHF

<table>
<thead>
<tr>
<th>Drug</th>
<th>Efficacy</th>
<th>Dosing/administration</th>
<th>Toxicity</th>
<th>Overall rating</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frusemide</td>
<td>++</td>
<td>Convenient. Oral/IV</td>
<td>↓Low Na &amp; K; otherwise safe</td>
<td>Component of standard combination therapy; used alone occasionally</td>
</tr>
<tr>
<td>Captopril</td>
<td>+++</td>
<td>Difficult dose administration in infants; oral route only</td>
<td>↓BP, ↓renal function,↑K, troublesome cough</td>
<td>Part of standard therapy; avoid in obstructive lesions</td>
</tr>
<tr>
<td>Digoxin</td>
<td>+++</td>
<td>Difficult dose calculation &amp; administration, especially IV. Oral &amp; IV.</td>
<td>Narrow safety margin; slow clearance; arrhythmogenic</td>
<td>Part of standard therapy. Use with extreme caution, especially if given IV</td>
</tr>
<tr>
<td>Dopamine/Dobutamine</td>
<td>++</td>
<td>Difficult dose calculation &amp; administration. IV drip only</td>
<td>Tachycardia; arrhythmogenic</td>
<td>Use only in critical cases (shock)</td>
</tr>
</tbody>
</table>
The most clinically demanding pediatric cardiac emergencies presentation

1.
2. Myocardial disease
3.
4.
5.
6.
CHF due to impaired myocardium – primary inotropic depression

- Myocarditis
- Cardiomyopathy
- Coronary perfusion abnormalities

Cardiogenic Shock
Management

1. Diuretic therapy
2. Inotropic support
   - Dopamine
   - Dobutamine: useful in impaired myocardial perfusion
3. After load reduction (ACE) inhibitors
4. Oxygen therapy
5. Digoxin
## Drugs for pediatric heart failure and the effects they produce

<table>
<thead>
<tr>
<th>Group</th>
<th>Drug</th>
<th>Effect</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>Digoxin, Diuretics, Captopril</td>
<td>↑Contractility, ↓heart rate, ↓Preload, anticongestive, ↓Afterload, ↓preload</td>
</tr>
<tr>
<td>B</td>
<td>Dopamine, Dobutamine</td>
<td>↑Heart rate, ↑contractility, ↑renal blood flow, ↑Contractility and rate</td>
</tr>
<tr>
<td>C</td>
<td>Carvedilol, Milrinone</td>
<td>↑Contractility, vasodilatation (Inodilator), ↑Contractility, vasodilatation (Inodilator)</td>
</tr>
<tr>
<td>D</td>
<td>Nesiritide, Levosimendan, Inhaled nitric oxide, Adenosine, Candesartan</td>
<td>Recombinant BNP: Natriuresis, diuresis, vasodilatation, Ca sensitizer: ↑contractility, vasodilatation (Inodilator), Pulmonary vasodilator, Pulmonary vasodilator; antiarrhythmic, Angiotension receptor blockade: vasodilatation</td>
</tr>
</tbody>
</table>

**Availability:**
- **Group A:** Widely available and widely used
- **Group B:** Widely available, but used in special circumstances
- **Group C:** Restricted availability
- **Group D:** Very restricted availability
The most clinically demanding pediatric cardiac emergencies presentation

1. Congestive heart failure
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4. Hypoxemic attack
5. Cardiac arrhythmias
6. Acute postoperative pulmonary hypertension (Pulmonary Hypertensive Crisis)
- Newborn with collapsed systemic circulation

OR

- Intense cyanosis

Emergency intervention is crucial
Newborn with obstructed flow

- CHF due to obstructive lesions is almost always associated with left sided lesions
  - Preductal coarctation of aorta
  - Hypoplastic left heart syndrome
  - Interrupted aortic arch

- Combination of
  - Shock
  - Pulmonary edema
  - Cardiogenic shock
Duct Dependant Circulation

1. Tricuspid Atresia
2. Hypoplastic Left Heart Syndrome
3. Pulmonary Atresia With Intact IVS
4. Pre-ductal Coarcotation
5. Interrupted Aortic Arch
6. Critical Aortic Stenosis
High blood pressure before point of coarctation

Low blood pressure beyond point of coarctation

Coarctation of the aorta
Evaluation of the infant’s pulses is the most important part of physical examination:

- Absent femoral pulses
Coarctation of the Aorta

Incidence 5% of CHD

Male: Female 2:1
Hypoplastic Left Heart Syndrome
Hypoplastic Left Heart Syndrome

Incidence  8%

Most common cause for early cardiac death

Presentation  first 1-3 weeks of age
Pulmonary Atresia
Management of Duct Dependant lesion

- Prostaglandin E1 (PGE1) is standard medical intervention
- Inotropic support need
- Ventilator support
- Emergency life saving surgery
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6. Acute post operative Pulmonary Hypertension (Pulmonary hypertensive crisis)
Hypoxemic Attack

Hyercyanotic spells → cyanotic spells

- Can occur in any cyanotic CHD with restricted pulmonary blood flow e.g.
  - Fallot’s tetralogy
  - Pulmonary atresia with VSD
  - Tricuspid atresia with restricted VSD
Cyanotic spell

Children with Tetralogy of Fallot exhibit bluish skin during episodes of crying or feeding.

“Tet spell”
Tetralogy of Fallot

Four abnormalities that result in insufficiently oxygenated blood pumped to the body:

1. Narrowing of the pulmonary valve
2. Thickening of the wall of the right ventricle
3. Displacement of the aorta over the ventricular septal defect
4. Ventricular septal defect—opening between the left and right ventricles
Tetralogy of Fallot X-ray
Pulmonary Atresia
Hypoxemic Attack

The pathophysiology of hypercyanotic spells is not entirely clear

Imbalance between pulmonary & systemic vascular resistance

- Spells do not occur during sleep
- Occur more frequently early in the morning or after a nap
Hypoxemic Attack

Precipitating factor:
- Fever
- Anxiety
- Exercise
- Dehydration
- Tachypnea

It is more common in the children with cyanosis and iron deficiency
Clinical Finding

- Sudden onset of labored deep breathing
- Either irritable and crying or lethargic and even unconscious
- Ashen or gray color
- Absent of previously heard murmur
- Drop in transcutaneous oxygen saturation
# Steps for the Management of Hyercyanotic spells

**Life Threatening Emergency**

<table>
<thead>
<tr>
<th>Medical management</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Calm the child and establish knee-chest position</td>
<td>The parent is the best person to calm the child</td>
</tr>
<tr>
<td>2. Oxygen</td>
<td>Blow-by O2</td>
</tr>
<tr>
<td>3. Sedation</td>
<td>Morphine: 0.1mg/kg IV, IM, or SQ ketamine: 1 mg/kg IV (Administer slowly as bolus infusion can cause respiratory arrest).</td>
</tr>
<tr>
<td>4. Establish intravenous access</td>
<td>Draw blood gas (usually capillary or venous). Draw arterial blood gas only if child is well – sedated.</td>
</tr>
<tr>
<td>5. Correct acidosis</td>
<td>Sodium bicarbonate 1 mEq/kg and repeat as necessary</td>
</tr>
<tr>
<td>6. Hydrate</td>
<td>10 cc/kg boluses of crystalloid or colloid</td>
</tr>
<tr>
<td>7. Pharmacological therapy:</td>
<td></td>
</tr>
<tr>
<td>- Propranolol</td>
<td>0.05 mg/kg/ IV. Maximum doe is 0.1mg/kg, not to exceed a total of 1mg. (Be prepared for severe bradycardia)</td>
</tr>
<tr>
<td>- Esmolol Infusion</td>
<td>Initial bolus of 0.5-1.0 mg/kg; then continuous infusion of 100-300 mcg/kg/min</td>
</tr>
<tr>
<td>8. General Anesthesia</td>
<td></td>
</tr>
<tr>
<td>9. Emergency cardiopulmonary bypass or ECMO or emergency surgery</td>
<td>Palliative or complete repair</td>
</tr>
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CHF due to Tachyarrhythmia

- Disturbances in cardiac rhythm are relatively common in infants, children

- Increase incidence of cardiac arrhythmias in children can be explained by:
  
  Advances in cardiac surgery that have resulted in survival of children with complex CHD
# Heart Rates in Children

## Infant

<table>
<thead>
<tr>
<th>Rate (BPM)</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>85</td>
<td>Normal</td>
</tr>
<tr>
<td>220</td>
<td>Sinus Tachycardia (SVT)</td>
</tr>
<tr>
<td>300</td>
<td>SVT</td>
</tr>
</tbody>
</table>

## Child

<table>
<thead>
<tr>
<th>Rate (BPM)</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>60</td>
<td>Normal</td>
</tr>
<tr>
<td>180</td>
<td>Sinus Tachycardia (SVT)</td>
</tr>
<tr>
<td>200</td>
<td>SVT</td>
</tr>
</tbody>
</table>
Conduction System

- Sinoatrial (SA) Node
- Atrioventricular (AV) Node
- Atrial Conduction
- Bundle of His
- Purkinje Fibers
- Right Bundle Branch
- Left Bundle Branch
Pathophysiology

- Most of children with rhythm disturbances
  No cause is recognizable
- Some CHD has relative high incidence of arrhythmia
  - Corrected TGA
  - Ebstein’s anomaly
  - Congenital mitral stenosis
- Post surgical repair of CHD:
  - TGA
  - Fallot’s tetralogy
  - Fontan procedure
- **Acquired heart disease**
  - Cardiomyopathies
  - Viral myocarditis
  - Cardiac tumors
  - Rheumatic carditis

- **Systemic disease:**
  - Electrolyte disturbances
  - Hyperthyroidism
  - Collagen disease (SLE)
  - Haemotologic disorders (thalassemia major)

- **Drug**
  - Theophylline
  - General anesthesia
  - Digitalis
## Features of Sinus Tachycardia and Supraventricular Tachycardia

<table>
<thead>
<tr>
<th>Feature</th>
<th>Sinus Tachycardia</th>
<th>Supraventricular tachycardia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart rate</td>
<td>Up to 240 bpm</td>
<td>200 – 300 bpm</td>
</tr>
<tr>
<td>Heart rate variation</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>P waves on ECG</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Response to adenosine</td>
<td>No response</td>
<td>Tachycardia stops abruptly</td>
</tr>
</tbody>
</table>
Clinical Manifestation

- Symptoms of CHF

- Symptoms related to decreased cerebral blood flow, syncope, irritability, dizziness.

- Palpitation
The PR interval is short and there is a slurred upstroke of the QRS complex noted in lead 2, AVF, and V2 through V6.
Wolff-Parkinson-White Syndrome
Long QT syndrome

Long QT is diagnosed because of a QTc of > 450 in a patient with syncope.
Long QT syndrome
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6. Acute postoperative pulmonary hypertension
   (Pulmonary Hypertensive Crisis)
• Pulmonary arterial hypertension can be defined as an increase in pulmonary arterial pressure (PA pressure) in the pulmonary vascular bed.

• PHTN is defined as a systolic pulmonary arterial pressure (PAP) of >35 mm Hg or a mean PAP of >25 mm Hg.

• In the clinical setting, the PAP is often described as a pressure relative to the systemic blood pressure.

• This measurement is determined by echocardiography most of the time.
Patients at particular risk for postoperative pulmonary hypertension can be identified preoperatively based on their cardiac disease and can be grouped into four broad categories based on the mechanisms responsible for pulmonary hypertension:

1) increased pulmonary vascular resistance

2) increased pulmonary blood flow with normal pulmonary vascular resistance

3) a combination of increased pulmonary vascular resistance and increased blood flow

4) increased pulmonary venous pressure.
The pathophysiology of acute PHTN in the postoperative period includes:

1) RV pressure overload and ventricular dysfunction.
2) Pulmonary blood flow decreases and RV pressure and volume increase.
3) The interventricular septum shifts toward the left ventricle.
4) Alters the compliance of the left ventricle
5) Increase in left ventricular (LV) end-diastolic pressure,
6) Increase in left atrial pressure, a decrease in pulmonary venous return,
7) Reduction in LV preload and cardiac output.

- **Pulmonary hypertensive crisis**
  Can escalate into a life-threatening event.
Hypotension and tachycardia are early signs of elevated PAP.

Bradycardia and the acute onset of pallor, resulting from a sudden fall in cardiac output, are ominous signs of a pulmonary hypertensive crisis and impending cardiac arrest.

The arterial oxygen saturation may remain in a normal range during a pulmonary hypertensive crisis unless there is intracardiac shunting, in which case a fall in arterial oxygen saturations will be an early sign of an acute increase in PAP.
Table 1. **Acute management of pulmonary hypertension**

**Reduce sympathetic stimulation**
- Maintain adequate sedation and analgesia; consider the use of muscle relaxants
- Premedicate before noxious stimuli, such as endotracheal tube suctioning
- Maintain normothermia

**Lower pulmonary vascular resistance**
- **Gas exchange**
  - ↑ Alveolar oxygen tension
  - Alkalosis/treat acidosis (metabolic or respiratory)
  - Hypocapnia (may compromise cerebral oxygenation)

**Mechanical ventilation**
- Maintain adequate functional residual capacity
- Avoid hypo- or hyperinflation
- Minimize intrathoracic pressure

**Vasodilating drugs**
- **Specific:** inhaled nitric oxide
  - Aerosolized iloprost
- **Nonspecific:** cGMP system: nitroprusside
  - cAMP system: phosphodiesterase type 3 inhibitors (milrinone)
    - Phosphodiesterase type 5 inhibitor (sildenafil)
    - Isoproterenol.
  - Prostacyclin I2, prostaglandin E1
Sildenafil has shown to work synergistically with NO, enhancing the efficacy of exogenous NO, which further increases vasodilatation.

This combined approach facilitates weaning from high doses of nitric oxide, by reducing the rebound effect commonly seen with discontinuation of inhaled NO.
5 months old admitted to LH ICU via ER on 16/11/2013 with severe respiratory distress and desaturation

He was markedly tachycardic, tachypneic
SPO2 84% on room air.
Birth history: Born in hospital weight 2.4kg
Discharged after routine vaccination.

Admitted to regional hospital at 15 days of age with picture of CCF, chest X ray showed cardiomegaly.

2D Echo. Showed: HLHS started on lasix and prostaglandin and shifted to SKMC.

In SKMC diagnosed as Unbalanced AVSD and discharged on captopril and lasix.

Readmitted to SKMC on 29/10/2013 at 4 months of age with increasing cyanosis.

Pre operative diagnosis: Unbalanced AVSD, RV dominant, moderate TR, mild MR, PDA, Patent foramen ovale, Pulmonary HT with R-L shunt across VSD.

Operated on 31/10/2013
Post op was on nitric oxide, sildinafil and anti-failure treatment.

Post op 2D Echo: Moderate RVH, LV volume markedly improved and RV dilatation resolved, mild TR, Tiny Residual VSD leak.

Discharged on 13/11/2013 on captopril and lasix
Chest X ray on admission

16/11/2013
LEFT ANTERIOR FASCICULAR BLOCK (QRS AXIS 15-45, OR IN I, RS IN III)
LEFT VENTRICULAR HYPERTROPHY WITH REPOLARIZATION ABNORMALITY (VOLTAGE CRITERIA PLUS ST/T ABNORMALITY)
ABNORMAL ECG

UNCONFIRMED REPORT

Site: 0 Cart: 0 Version 14100 Sequence #09005 25mm/s 10mm/mV 0.05-150 Hz
Management

- Respiratory support with CPAP
- Iv fluids
- Iv Ceftrioxone
- He developed sudden cardio respiratory arrest two hours after ICU admission.
- Immediately revived with CPR, intubated and ventilated.
• Post arrest had hypotension and hypoglycaemia, managed with normal saline bolus, iv dextrose and dobutamine infusion.

• 2D Echocardiogram: RA, LA dilated, R-L shunt at atrial level, small residual VSD shunt, TR V\text{max} 5.3 m/s, severe pulmonary hypertension (Pulmonary hypertensive crisis).
• Started on nitric oxide.
• Other supportive measures, FFP, vitamin k, packed RBC, Calcium gluconate, Lasix and captopril continued
• Repeat 2D Echo next day: TR $V_{\text{max}}$ 4.9 M/S
• Started on sildenafil and increased gradually.
• Repeat 2D Echo on 24/11/2013: mild to mod TR, mild to mod MR, L–R shunt at atrial level.
Nitric oxide gradually weaned and stopped in 5 days.

Remained on low ventilator parameters.

Dobutamine stopped within one week.

Extubated after 12 days

Continued on Lasix, captopril and sildenafil and discharged in good condition on 4/12/2013.