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# Heart Failure

Standard Operating Procedure (SOP)



*Union of Myanmar Special region - 1*  
*Pediatric (SOP)*

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## Heart Failure

A pathologic condition in which the heart is unable to pump sufficient blood to meet the metabolic demands of the body, As the demands on the heart outstrip the normal range of physiologic compensatory mechanisms, signs of congestive heart failure occur.

*Diagnosis*

*History*

- Acute myocarditis with a recent history suggestive of acute viral infection
- History of previously diagnosed congenital or rheumatic heart disease, history suggestive of rheumatic fever
- History of hypertension and suggestive history of acute or chronic renal diseases
- History of other medical diseases e.g Thalassaemia, Malignancy, Thyrotoxicosis, Anemia
- History of similar attack and hospitalization before
- History of taking any medications (e.g. Cytotoxics especially Adriamycin)

*Clinical Presentation*

*Clinical manifestation of congestive heart failure vary with the age of the child.*

**Table 3.1 Symptoms characteristic of heart failure in children**

|                                       | <i>Commonly encountered</i>                                                                                                                                                               | <i>Less commonly encountered</i>                                                                                                                                       |
|---------------------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <i>Infants and young children</i>     | <ul style="list-style-type: none"><li>• Tachypnea</li><li>• Feeding difficulty (reflux, vomiting, feeding refusal)</li><li>• Diaphoresis</li><li>• Pallor</li></ul>                       | <ul style="list-style-type: none"><li>• Cyanosis</li><li>• Palpitations</li><li>• Syncope</li><li>• Facial edema</li><li>• Dependent edema</li><li>• Ascites</li></ul> |
| <i>Older children and adolescents</i> | <ul style="list-style-type: none"><li>• Fatigue</li><li>• Effort intolerance</li><li>• Dyspnea</li><li>• Orthopnea</li><li>• Abdominal pain</li><li>• Nausea</li><li>• Vomiting</li></ul> | <ul style="list-style-type: none"><li>• Palpitations</li><li>• Chest pain</li><li>• Dependent edema</li><li>• Ascites</li></ul>                                        |

*Source: Kantor PF et al, Presentation, Diagnosis and Medical management of Heart Failure in Children.*

*Canadian Cardiovascular society Guideline, Canadian Journal of Cardiology 2013; (29): 1537.*

## Examination

### Signs of reduced tissue perfusion (Low cardiac output)

- Weakness, pallor
- Peripheral cyanosis with cold clammy skin
- Lowr blood pressure, weak pulse, oliguria
- Cardiogenic shock

### Signs of pulmonary venous congestion

- Tachypnea, respiratory distress (retractions)
- Grunting, difficulty with feeding
- Rales, pulmonary oedema

### Signs of right-sided venous congestion

- Hepatomegaly
- Oedema or ascites
- Jugular venous distention (It is not a reliable indicator in infants)

Uncompensated congestive heart failure in an infant primarily manifests as a failure to thrive. in severe cases, failure to thrive may be followed by signs of renal and hepatic failure.

- Absence of murmur could not exclude underlying heart diseases
- Assessment of the upper and lower limbs blood pressures and femoral pulses are crucial
- Left ventricular failure and pulmonary edema are not always synonymous as there are other causes for pulmonary edema e.g. acute renal failure, acute respiratory distress syndrome (ARDS)

The New York Heart Association (NYHA)/Ross classification can be used in pediatric heart failure and is helpful to guide medical management but is not to determine the prognosis.

### Table 3.2 Classification of symptomatic severity in pediatric heart failure

| Class of symptoms | Symptoms noted on history                                                                                                |                                                                                       |
|-------------------|--------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------|
|                   | Infants                                                                                                                  | Older children                                                                        |
| I                 | Asymptomatic                                                                                                             |                                                                                       |
| II                | <ul style="list-style-type: none"><li>• Mild tachypnea or diaphoresis with feeding</li><li>• No growth failure</li></ul> | <ul style="list-style-type: none"><li>• Dyspnea on moderate exertion</li></ul>        |
| III               | <ul style="list-style-type: none"><li>• Marked tachypnea or diaphoresis with feeding</li><li>• Growth failure</li></ul>  | <ul style="list-style-type: none"><li>• Dyspnea on mild or minimal exertion</li></ul> |
| IV                | <ul style="list-style-type: none"><li>• Tachypnea, diaphoresis or respiratory distress at rest</li></ul>                 |                                                                                       |

Source: Kantor pf et al. Presentation, Diagnosis and Medical management of Heart Failure in Children. Canadian Cardiovascular Society Guideline, Canadian Journal of Cardiology 2013; (29): 1538.

## Investigations

### Chest x-ray

- Size of heart- Enlarged (LVH or RVH or BVH)
  - Pulmonary vascularity - Oligaemic or plethoric
  - Pulmonary conus- Enlarged  $\pm$
  - Pulmonary oedema: the lungs will appear whiter often with a typical 'bats wing' distribution of oedema, dilated pulmonary capillaries and upper lobe diversion
  - Chest X-ray allows the differential diagnosis of left heart failure from inflammatory or infectious lung diseases
2. **A 12-lead ECG** may reveal evidence of structural disease (RAD or LAD, LVH or RVH or BVH, RsR' pattern  $\pm$ , Strain pattern +) or a complete atrioventricular block or arrhythmia
  3. **Echocardiography** is indicated to assess cardiac function and identify potential cardiovascular causes, particularly structural lesions
  4. **Laboratory testing**
    - Oxygen saturation
    - FBC count and hemoglobin concentration
    - Urea, creatinine level and electrolytes
    - Calcium level
    - Hepatic function
    - BNP/NT pro BNP level (if available) is useful in distinguishing heart failure from respiratory or other non-cardiac disease and also serial BNP levels are useful in monitoring response to treatment
  5. **Pulse oximetry**, test in newborns may be useful. The significant increased in oxygen saturation when the patient is receiving 100% oxygen may help indistinguishing intracardiac mixing malformations from pulmonary disease in the setting of hypoxia

## Management

As the causes of congestive heart failure vary and they appear in variable degrees. the medical management should be tailored to the specific details of each case.

Acute congestive heart failure in the neonate or infant

- Immediate concern - potential sepsis or ductal-dependent congenital heart disease
- Assessment of the patient's airway, breathing, and circulation (ABCs)
- Admission to the Neonatal ICU

- *Laboratory testing, blood culture and empiric antibiotic therapy*
  - *Management of low cardiac output can be initiated by*
    - *Dopamine infusion at 5-10 ug/kg/min*
    - *Acidosis can be corrected with the administration of fluid and/or bicarbonate*
    - *Calcium should be administered when hypocalcemia is documented*
  - *Echocardiography should be considered. Conditions to consider in young infants with a more protracted congestive heart failure course include*
    - *Significant left-to-right shunts (including large ventricular septal defects aortopulmonary shunts and arteriovenous malformations)*
    - *Ductal-dependent structural heart disease (coarctation or interruption of the aortic arch, total anomalous pulmonary venous return, hypoplastic left heart syndrome, truncus arteriosus, pulmonary atresia and transposition of the great arteries)*
- Absent femoral pulses suggests a ductal-dependent lesion, and treatment with alprostadil (PGE1) is warranted.*

## **Acute heart failure in the older child**

### **In heart failure with reduced tissue perfusion (Low Cardiac Output)**

- *Children presenting with heart failure because of reduced cardiac output with end-organ dysfunction are likely to benefit from inotropic therapy as a rescue strategy.*
- *Dopamine infusion (5-10ug/kg/min) is first line inotropic agent.*

#### *In heart failure with severe hypertension*

- *Left ventricular function is usually preserved and patients should be treated with vasodilators.*
- *Most children with hypertensive heart failure have chronic or acute renal disease and careful attention to fluid balance and diuresis is essential.*
- *Furosemide is a drug of choice in acute glomerulonephritis with fluid overload.*

### **In heart failure with signs of pulmonary venous congestion**

- *Left ventricular function is usually preserved and patients should be treated mainly with diuretics and if required, bronchodilators and narcotics; Morphine should be added.*
- *Respiratory support may be necessary with continuous positive airway pressure (CPAP) or non-invasive positive pressure ventilation may be required.*

### **In heart failure with signs of right-sided venous congestion**

- *Fluid overload is managed mainly with diuretics.*

*Underlying causes must be treated with specific agents.*

*Indiscriminate administration of intravenous fluid resuscitation is contraindicated if there is no reason to achieve an optimal preload and it will worsen the condition.*

- *In reduced tissue perfusion: Inotropic agents*
- *In hypertensive heart failure: IV Vasodilator +/- IV Diuretic (Sublingual Nifedipine is contraindicated)*
- *In pulmonary venous congestion: IV Diuretic +/- Vasodilator +/- Narcotics*
- *In both reduced tissue perfusion and pulmonary venous congestion: Inotropic agents + IV Diuretic*
- *In right-sided venous congestion: IV Diuretic*

## Inotropic Agents

- *Inotropic therapy should be confined to patients with depressed systolic function and clinical evidence of low cardiac output syndrome who can be closely monitored for tachyarrhythmias and blood pressure lability.*
- *In this setting, dopamine, dobutamine, and low dose epinephrine have all shown efficacy in children.*
  - *Dopamine (5-10 ug/kg/min) to augment contractility until stabilization is achieved*

## How to prepare dopamine infusion

$$\frac{\text{mg of dopamine in 100 ml}}{(\text{N/S or D/S})} = \frac{6 \times \text{body weight (kg)} \times \text{Dose (ug/kg/min)}}{\text{Rate (m/hr)}}$$

*Dopamine is more effective than dobutamine for short term treatment (including preterm infants).*

*If the requirement of Dopamine is >10 µg/kg/min, Dobutamine should be added*

- *Dobutamine (5-10 ug /kg/min)*
    - *Significant positive inotropic effect and mild peripheral vasodilatation and (so should be avoided in patient with moderate or severe hypotension.*
  - *Epinephrine (Adrenaline)*
    - *It is used for severe heart failure refractory to dopamine. It should be started at a dose of 0.05 µg/kg/min and titrated to maximum 1.0 µg.kg/min.*
    - *In severe cases, patients may receive doses of 2-3 ug/kg/min IV or even higher.*
- NB: At high doses, extremities become ischemic and even turn dark and potentially necrotic. Ventricular dysrhythmias may be precipitated, Rapid IV infusions may cause death from cerebrovascular hemorrhage or cardiac arrhythmias.*

## Diuretics

- *They help by reducing circulatory volume and thereby reducing preload.*
- *A loop diuretic, such as furosemide, is recommended for patients with signs and symptoms of congestion.*
- *An initial starting dose of 0.5-1 mg/kg intravenously or orally every 6-12hours, is safe and effective.*

- *Thiazide diuretic agents (2 mg/kg/ day) will significantly enhance the effect of loop diuretic agents and can be added in patients with limited response to loop diuretic agents.*
- *Hypokalemia or hyponatremia are commonly noted with this combination and therefore electrolyte monitoring is important.*

## Morphine

- *Morphine 0.05-0.1 mg/kg IV over 5 min or 0.1-0.2 mg/kg SC reduces the terror and anxiety and it also reduces preload and afterload by venodilatation and mild arterial dilatation.*
- *It is indicated in a patient with severe heart failure especially associated with restlessness and dyspnoea.*

## Vasodilator agents

- *They are recommended mainly in patients with hypertensive heart failure to reduce afterload and preload.*
- *They might have an adjunctive role in stabilizing patients with reduced systolic function and associated hypertension because of renal dysfunction or other causes.*
  - *IV Hydralazine (arterial vasodilator) - 0.1-0.4 mg /kg (Maximum 10 mg) slow IV >3-5 minutes. It can repeat every 4-6 hours or continuous infusion 4-6 ug/kg/min (Maximum 300 ug/min). It is contraindicated in high intracranial pressure, cerebrovascular diseases, severe renal impairment mitral valvular heart diseases and hypertrophic cardiomyopathy.*
  - *Glyceryl Trinitrate: It is a predominantly venodilator and an IV infusion may be useful in reducing preload and afterload and may also improve coronary blood flow, Initiate with 1.0 ug/kg/min and titrate 1.0-10 ug/kg/min (Maximum-400 ug/min)*
- *Other alternatives to hypertensive heart failure*
  - *Nicardipine - Continuous IV infusion 1.0-3.0 ug/kg/min (Maximum - 0250 ug/min), it is similar to Nifedipine without the deleterious risk of depressing myocardial function. It is contraindicated in high intracranial pressure, cerebrovascular diseases.*
  - *Diazoxide (arterial vasodilator) - 1-3 mg/kg dose.*
  - *Nitroprusside Sodium Arterial vasodilator)-0.3-8.0 ug/kg/min infusion. Maximum final concentration is 200 µg/min, If maximum dose is used for >10 min without good blood pressure control, stop it because of cyanide toxicity risk, It is contraindicated in coarctation of aorta, high intracranial pressure and intracranial hemorrhage.*

### Contraindication to Hypertensive heart failure

- *Labetalol (alpha and beta blocker) is contraindicated in heart failure.*
- *Sublingual Nifedipine is not recommended as it may cause a precipitous drop in blood pressure particularly in those with long standing hypertension (or uncertain duration).*

## Chronic Heart Failure

*If the underlying cause of the congestive heart failure cannot be immediately corrected in a patient who is hemodynamically stable, management can be initiated by using several agents.*

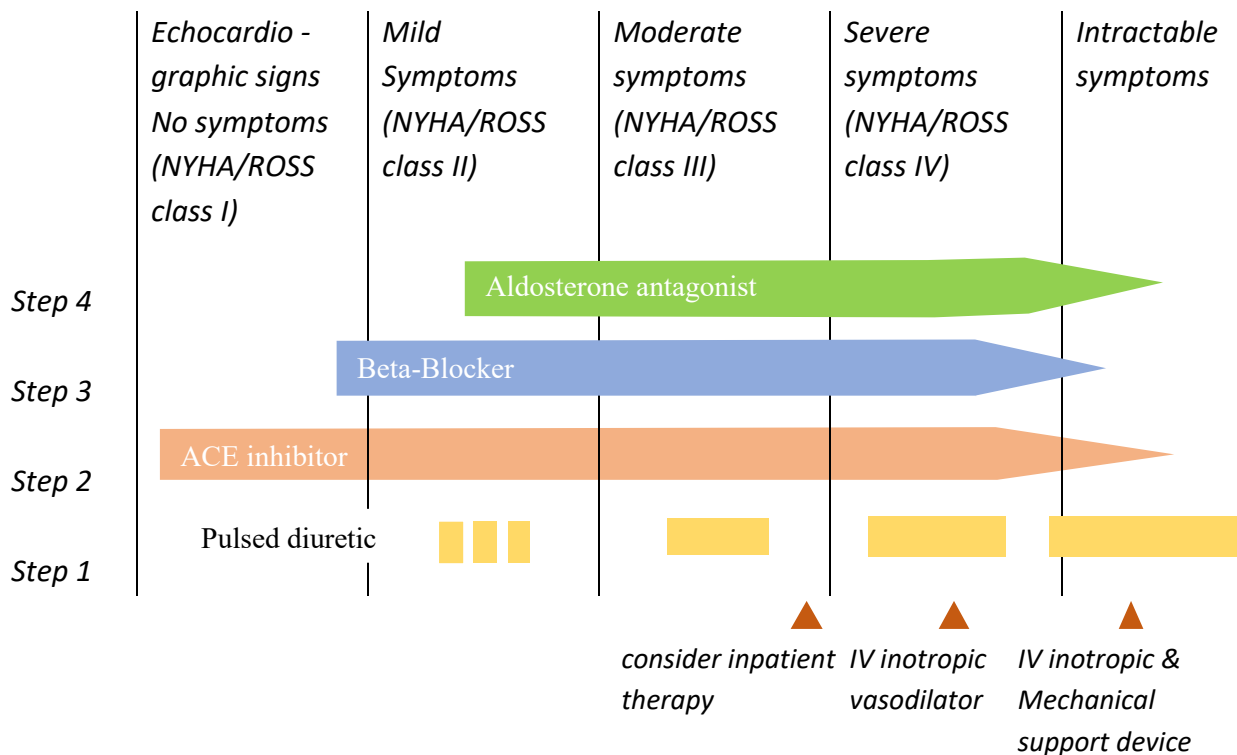
- **Angiotensin Converting Enzyme Inhibitors (ACEI)**
  - *They are the medications of choice in patients who have*
  - *Large left-to-right shunts at the ventricular or arterial level (VSD, ASD, PDA)*
  - *Left-sided regurgitant lesions (aortic or mitral regurgitation)*
  - *Left ventricular dysfunction (myocarditis or dilated cardiomyopathy)*
- *They are contraindicated in unrelieved outflow obstruction (MS, AS Coarctation, PS), Pink TOF and renal dysfunction.*
- *In advanced heart failure, ACEI introduction should occur after stabilization of heart failure symptoms and simultaneous to inotropic support withdrawal.*
- *Captopril is the first choice for most infants and enalapril for children older than 2 years.*
- *Caution is advised when these are used in the first 4 months of life because renal dysfunction is more common.*
- *In patients who are receiving diuretics, ACEI should be introduced cautiously with careful monitoring of blood pressure and urine output.*
- *Check urea and electrolytes before and after giving ACEI, every few months.*
- *A creatinine rise of greater than 50% over baseline value in any patient requires a reassessment of fluid balance and diuretic therapy and consideration for a dosage reduction or withdrawal of ACEI therapy.*
- *Spironolactone and supplemental potassium should be avoided except in the presence of documented hypokalemia.*
- *A small drop in systolic blood pressure is typically noted in patients who take an ACEI, This might occasionally exceed the expected 5%-10% drop in baseline values necessitating observation for up to 2 hours after the first dose.*
  - *Enalapril: test dose 0.05 mg/kg, increase as tolerated to 0.2-0.3 mg/kg OD at night time.*
  - *Captopril: 0.1-0.5 mg/kg/d divided 8 hourly*
  - *Enalapril: test dose 0.05 mg/kg, increase as tolerated to 0.2-0.3 mg/kg*
- **Angiotensin Receptor Blockers (ARB):** *Alternatively, losartan (0.1-0.5 mg/kg/day OD), may be used in patients in whom ACEI side effects (particularly cough) may be unacceptable.*
- **$\beta$ -Blockers**
  - *They counter the activation of the sympathetic nervous system which is often seen in heart failure.*
  - *The beneficial effects of  $\beta$  blocker in heart failure may include*
    - *Slowed heart rate*
    - *Improving balance of oxygen supply and demand*
    - *Reduced myocardial apoptosis and fibrosis*
    - *Antiarrhythmic effects*

- Synergism with ACE inhibitors
- Initiation of  $\beta$  blocker therapy might be considered in children who have persistent systolic dysfunction despite being established with ACEI.
- Carvedilol (a nonselective  $\beta$ -adrenoreceptor antagonist with  $\alpha$ -adrenergic blocking activity) is commonly used in children.
  - A starting dose of 0.05 mg/kg every 12 hour is required and in less than 4 years old, every 8 hours.
  - Adolescents - 1 mg/kg/day
  - Aged 2-11 years - 2 mg/kg/day
  - Aged 28 days to 23 months - 3 mg/kg/day
- Aldosterone antagonist

As it reduces some of the effects of RAA system activation, it reduces sodium and water retention, reduced myocardial fibrosis. It enhances the effect to loop and thiazide diuretics.

  - Spironolactone can be initiated for patients in whom therapy with ACEI and  $\beta$ -blocker has not resulted in improved ventricular function.
  - Close monitoring of renal function and potassium level is essential.
  - Dose - 1-2 mg/kg/day

## Stepwise introduction of medical therapy in heart failure



Indicates declining range of candidates for introduction

Indicates requirement for reduced or slowed up-titration

Figure 3.1. A symptom-based guide to the introduction of oral maintenance therapy for children with chronic heart failure. Lighter shading for each drug indicates when the possibility of a lower target dose or slower increment than usual is needed, because of advanced heart failure with severely compromised systolic function. Narrowing of the bars indicates the possibility that fewer patients will tolerate the introduction of that agent at that given symptomatic stage. Note the preferred use of pulsed diuretic therapy which becomes more frequent as symptoms advance. Red arrows indicate the point at which admission to hospital and additional support therapies might be required.

Source: Kantor PF et al. Presentation, Diagnosis and Medical management of Heart Failure in Children. Canadian Cardiovascular Society Guideline, Canadian Journal of Cardiology 2013; (29) 1546.

## Treatment of underlying cause

- Rheumatic Carditis
- Myocarditis
- Hypertension
- SVT
- Surgery in correctable lesions

## Management of Electrolytes

### Hyponatremia

- *It is caused by water retention in response to vasopressin but may be exacerbated by use of diuretics and salt wasting by the kidney.*
- *Sodium supplementation is never indicated except in emergency situations. Severe hyponatremia is generally best managed by reducing the dose of diuretics or by restricting fluid intake.*
- *In any child taking more than 2 mg/kg/d of furosemide, electrolyte levels should be checked every few months or so as in any child taking furosemide in conjunction with other diuretics or ACEI.*

### Hyperkalaemia

- *Elevated potassium levels may represent renal compromise or even tissue destruction due to low cardiac output.*

### Hypocalcemia

- *Calcium should be administered when hypocalcemia is documented. Dose-10-20 mg/kg (0.1-0.2 ml/kg of calcium chloride 10%) IV administered at an infusion rate that does not exceed 100 mg/min*

## General Treatment

### Oxygen and ventilatory assistance

- *The maintenance of an oxygen saturation within the normal range (95-98%) in hypoxaemic patients is important in order to maximize oxygen delivery to the tissues and thus helping to prevent end organ dysfunction and multiple organ failure.*
- *Hyperoxia can be associated with reduced coronary blood flow, reduced cardiac output, increased systemic vascular resistance and a trend to higher mortality.*
- *The use of increased concentrations of oxygen in patients without evidence of hypoxaemia is more controversial and may cause harm.*

### Infections

- *Patients with advanced heart failure are prone to infectious complications, commonly respiratory tract infections, urinary tract infections, septicaemia, or nosocomial infection.*
- *Fever may be absent, Meticulous infection control and prompt antibiotic therapy should be given.*

### Acidosis

- *Routine administration of sodium bicarbonate is not recommended because bicarbonate may worsen intracellular acidosis and may result in a decrease in myocardial contractility.*
- *Acidosis should be corrected with increased perfusion from use of cardiotropic medications with optimal ventilation.*

*Fever: Fever should be treated aggressively as fever can precipitate heart failure*

*Anemia: Correct anemia as it aggravates congestive heart failure by increasing the demands for cardiac output.*

*Nutrition: Failure to thrive is an indication for increased medical management or surgical repair of structural heart disease.*