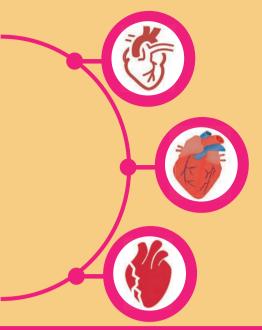




Standard Treatment Workflow (STW)

PEDIATRIC HEART FAILURE

ICD-10-150.9



DEFINITION

Clinical and pathophysiological syndrome that results from inability of the heart to function adequately to meet the metabolic demands of the body

CLINICAL SPECTRUM

- · Acute decompensated HF
- · Chronic compensated HF
- · Acute exacerbation of chronic HF

MODIFIED ROSS CLASSIFICATION OF HEART FAILURE

- · Class I: No symptoms/limitations
- · Class II: Mild tachypnea/sweating during feeds in infants/ dyspnoea on exertion in older children but no growth failure
- Class III: Significant tachypnea or sweating during feeds/marked dyspnoea on exertion/prolonged feeding time with growth failure
- · Class IV: Symptoms (tachypnoea, retractions, grunting and sweating) even at rest with growth failure

HEART FAILURE OFTEN HAS A TREATABLE CAUSE IN MOST CHILDREN. IDENTIFYING AND TREATING THE CAUSE IS THEREFORE THE MOST IMPORTANT PRIORITY

| Category | Specific Conditions |
|---------------------|-------------------------------------|
| Shunt lesions | VSD, PDA, AP window, AVCD, TGA, |
| | Truncus, TAPVC |
| Obstructive lesions | Critical AS, PS, coarctation/aortic |
| | interruption |
| Regurgitant lesions | Congenital- AV canal defect, |
| | Ebsteins anomaly |
| | Acquired- RHD, IE, post-operative |
| Primary Myocardial | Dilated cardiomyopathy, Inborn |
| dysfunction | errors of metabolism, muscular |
| | dystrophy, drug induced |
| | |

| Category | Specific Conditions | |
|-----------------------|---------------------------------------|--|
| Inflammatory | Myocarditis and other | |
| | immunoinflammatory conditions | |
| Abnormal rate/rhythm | Tachycardiomyopathy, bradycardia, | |
| | AV dyssynchrony | |
| Ischemic | Anomalous coronary artery from | |
| | pulmonary artery, Coronary artery | |
| | occlusion from other causes | |
| Post- cardiac surgery | Variety of causes (cardiopulmonary | |
| | bypass, Myocardial preservation etc.) | |
| Abnormal homeostasis | Hypoxia, hypocalcemia, | |
| | hypoglycemia, sepsis, hypothermia | |
| | | |

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| | | |

- Duct dependent systemic circulation
 - HLHS
 - Critical AS
 - Critical Co A Interrupted arch
- Severe Tricuspid regurgitation
- Vein of Galen malformation
- · Fetal/Neonatal myocarditis
- · Congenital MR

7-30 Days

- VSD with Coarctation
- Large AP window
- Persistent truncus arteriosus
- Single ventricle physiology with no PS
- · TGA-VSD/PDA
- · Large VSD or PDA especially in preterm infants
- · All cases listed for the first week

3-6 Months

- Large post tricuspid L-R shunts
 - VSD
 - PDA
 - AV canal defects
- · ALCAPA
- Mvocarditis/DCM
- All examples listed for the 7-30 days category

6 Months - 1 Years

- Large post tricuspid L-R shunts
 - VSD
 - PDA
 - AV canal defect
- Mvocarditis/DCM
- · ALCAPA

1-10 Years Heart valve

- disease (RHD) Myocarditis/DCM
- Aortoarteritis · Palliated CHD
- · Post KD coronary arteriopathy
- · Idiopathic PAH

SYMPTOMS

Neonate

- Lethargy Fast breathing
- Poor suck
- Reduced urine output
- Cold extremities

Infant

· Rapid and labored breathing

- · Excessive sweating
- Feeding difficulties (suck-rest-suck cycles) · Puffiness of face,
- · Poor growth
- Frequent chest infections

Older children

- Breathlessness
- · Effort intolerance
- Growth retardation
- extremities
- Abdominal distension

SIGNS

- · Tachypnea and labored respiratory efforts with intercostal and subcostal recession (RR>60/min in less than 1 year old and >50/min in 1-2 year old)
- Tachycardia (HR>160/min in less than 1 year old, >140/min between 1-2 year old)
- Hepatomegaly
- · Auscultation-Crackles at lung bases (limited sensitivity and specificity)
- · S3 gallop, murmurs
- · Raised JVP (not useful in infants)
- · Peripheral edema

RED FLAGS

- Reduced peripheral perfusion
- Reduced urine output
- Elevated lactate levels
- Altered sensorium

INVESTIGATIONS

HEART FAILURE MIMICS

- Sepsis
- · Respiratory distress syndrome
- · Inborn errors of metabolism
- Bronchiolitis (infants)

ESSENTIAL INVESTIGATIONS

Chest x-ray

Information on cardiac silhouette, pulmonary vasculature, pulmonary artery dilatation and associated skeletal abnormalities

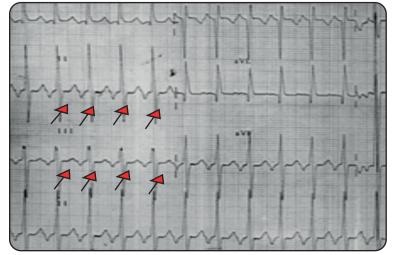
ECG

Diagnosis of treatable causes of heart failure such as persistent tachyarrhythmia, ALCAPA and, hypocalcemia. Other specific causes such as Pompe's disease, specific forms of cardiac muscle involvement in muscular dystrophy have ECG manifestations

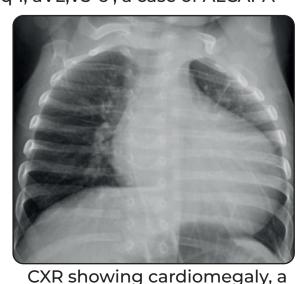
Echocardiogram

Critically important to accurate diagnosis and tailoring response to therapy

12 lead ECG showing classical pattern of q 1, aVL, V5-6, a case of ALCAPA



Tachycardiomyopathy is suggested by abnormal P waves (inverted in II, III and aVF) additional clues are fixed and rapid heart rates



case of dilated cardiomyopathy



Standard Treatment Workflow (STW)

PEDIATRIC HEART FAILURE

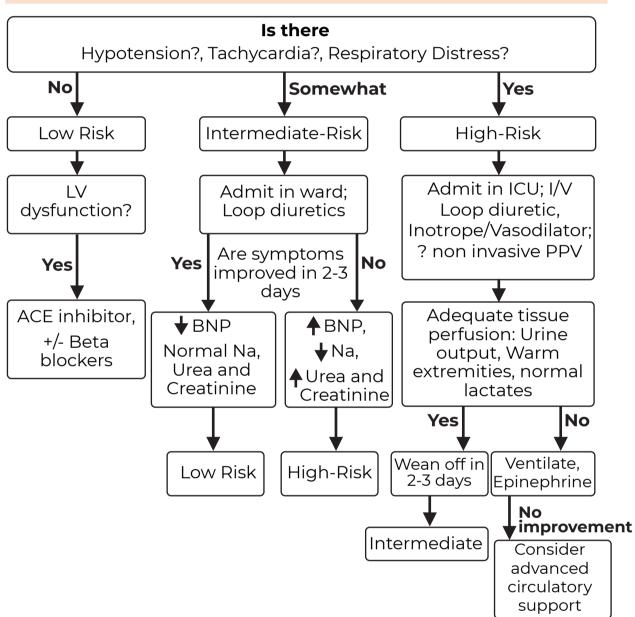
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| Essential blood tests to be performed in all | Utility | | |
|--|--|--|--|
| Complete blood count; CRP | Identifying Sepsis, Anemia | | |
| Electrolytes and urea, creatinine | Elevated urea, creatinine may indicate decompensated HF or may result from medication side effects. Electrolyte imbalance is a common association of HF and diuretic use. Hypocalcemia can cause ventricular dysfunction leading to HF | | |
| Liver function test | Elevated bilirubin, liver enzymes and prolonged prothrombin time points towards congestive hepatopathy. Hypoalbuminemia points to chronic HF and poor nutrition | | |
| Optional tests to be decided based on clinical situation | | | |
| Arterial blood gas with lactate | Lactic acidosis- as a marker of tissue perfusion and helps monitor response to treatment; It is also elevated in specific inborn errors of metabolism | | |
| Thyroid function test | Thyroid hormone imbalance could be a primary cause or may lead to worsening of symptoms | | |
| Brain Natriuretic Peptide (BNP) | It helps differentiate HF from respiratory disease. Useful in monitoring response to therapy | | |
| Cardiac enzymes (troponin I, T, CKMB) and Viral Panel | In suspected cases of myocarditis | | |

Management Goals

Correct the underlying cause Reduce associated morbidity and mortality

Improve functional status and quality of life



General Measures

Fluid restriction

 In acute HF with lung congestion, peripheral edema despite diuretics and in presence of hyponatremia

Rest and restriction of activity

 Activity as tolerated for older children with chronic compensated HF

Correction of Anaemia

 Hematinics; Blood transfusion only for severe anemia (Hb < 7gm/dl)

Nutrition

- NG feeds for infants in acute severe HF.
- In infants calorie intake of 120-150kcal/kg/with a fluid intake of 100 ml/kg/day. (thickening of feeds or by adding coconut oil/medium chain triglyceride). In older children increase protein content of diet while optimizing the fat and carbohydrate intake. Supplement Ca and Vit D3;
- Dietary restriction of sodium is generally not recommended in children unless there is severe edema unresponsive to diuretic therapy

Supplementary oxygen

 May be necessary when there is respiratory distress but must be used with caution in L-R shunts and avoided in neonates with duct dependent lesions

Inotropes should be physiologically appropriate:

- · Avoid vasodilators in presence of fixed outflow obstruction (AS); use vasodilators for regurgitant lesions, pump failure and large shunts
- · Avoid using very high doses for sustained periods (Preferably adrenaline < 0.1; dopamine or dobutamine < 15 mcg/g/min)

ABBREVATIONS

ACEI: Angiotensin Converting Enzyme Inhibitor

ALCAPA: Anomalous Origin of Left Coronary Artery from

Pulmonary Artery

AP Window: Aorto-Pulmonary Window

AS: Aortic Stenosis

AVCD: Atrio-Ventricular Canal Defect **AVCD:** Atrio-Ventricular Canal Defect

CoA: Coarctation of the Aorta

CKMB: Creatine Kinase Myoglobin Binding

CRP: C-reactive Protein **DCM:** Dilated Cardiomyopathy

HF: Heart Failure

HLH: Hypoplastic Left Heart

HR: Heart Rate

IE: Infective Endocarditis

JVP: Jugular Venous Pressure

KD: Kawasaki Disease

LV: Left Ventricle

MR: Mitral Regurgitation

NG: Naso-Gastric

PAH: Pulmonary Arterial Hypertension

TAPVC: Total Anomalous Pulmonary Venous Connection

PDA: Patent Ductus Arteriosus **PPV:** Positive Pressure Ventilation

PS: Pulmonary Stenosis

RHD: Rheumatic Heart Disease

RR: Respiratory Rate

TGA: Transposition of Great Arteries **VSD:** Ventricular Septal Defect

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