STANDARD TREATMENT WORKFLOW (STW)

PEDIATRIC HEART FAILURE

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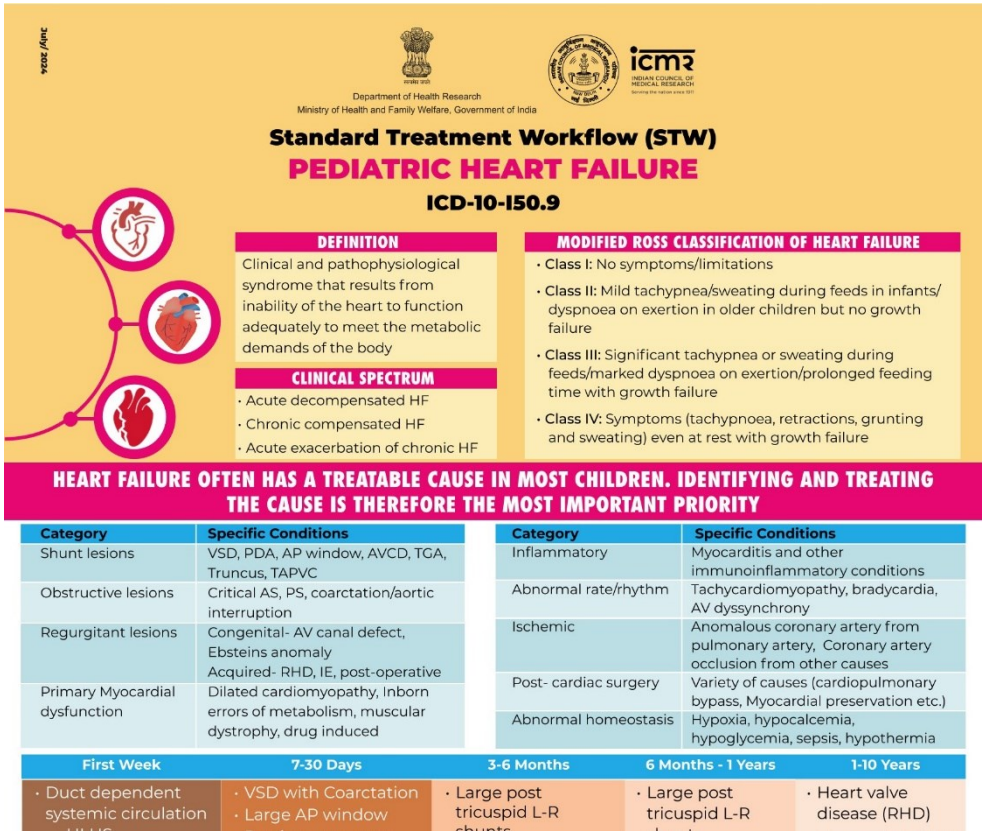

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Department of Health Research
Ministry of Health and Family Welfare, Government of India

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Standard Treatment Workflow (STW)

PEDIATRIC HEART FAILURE

ICD-10-I50.9

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 dysfunction	Dilated cardiomyopathy, Inborn 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

First Week	7-30 Days	3-6 Months	6 Months - 1 Years	1-10 Years
• Duct dependent systemic circulation ◦ HLHS ◦ Critical AS ◦ Critical Co A ◦ Interrupted arch • Severe Tricuspid regurgitation • Vein of Galen malformation • Fetal/Neonatal myocarditis • Congenital MR	• 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	• Large post tricuspid L-R shunts ◦ VSD ◦ PDA ◦ AV canal defects • ALCAPA • Myocarditis/DCM • All examples listed for the 7-30 days category	• Large post tricuspid L-R shunts ◦ VSD ◦ PDA ◦ AV canal defect • Myocarditis/DCM • ALCAPA	• Heart valve disease (RHD) • Myocarditis/DCM • Aortoarteritis • Palliated CHD • Post KD coronary arteriopathy • Idiopathic PAH

SYMPTOMS

Neonate	Infant	Older children
• Lethargy • Fast breathing • Poor suck • Reduced urine output • Cold extremities	• Rapid and labored breathing • Excessive sweating • Feeding difficulties (suck-rest-suck cycles) • Poor growth • Frequent chest infections	• Breathlessness • Effort intolerance • Growth retardation • Puffiness of face, 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

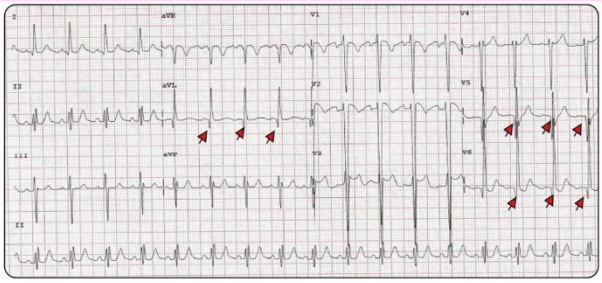
Information on cardiac silhouette, pulmonary vasculature, pulmonary artery dilatation and associated skeletal abnormalities

ECG

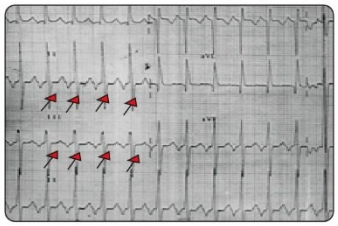
Diagnosis of treatable causes of heart failure such as persistent tachyarrhythmia, ALCAPA and, hypocalcemia. Other specific causes such as Pompe's disease, specific forms of cardiac muscle involvement in muscular dystrophy have ECG manifestations

Echocardiogram

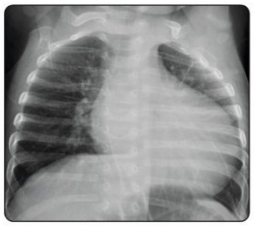
Critically important to accurate diagnosis and tailoring response to therapy



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



CXR showing cardiomegaly, a case of dilated cardiomyopathy

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(Continued)

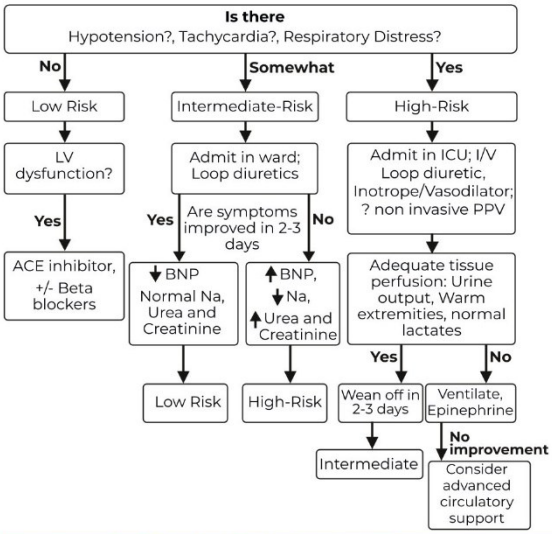
Essential blood tests to be performed in all	Utility
Complete blood count; CRP	Identifying Sepsis, Anemia
Electrolytes and urea, creatinine	Elevated urea, creatinine may indicate decompensated HF or may result from medication side effects. Electrolyte imbalance is a common association of HF and diuretic 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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PEDIATRIC HEART FAILURE IS BEST MANAGED IN CONSULTATION WITH A PEDIATRIC CARDIOLOGIST