July/ 2024

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Ministry of Health and Family Welfare, Government of India

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Standard Treatment Workflow (STW)

ACUTE RHEUMATIC FEVER

ICD-10-I01.9

Rheumatic fever (RF) is an acute, nonsuppurative inflammatory disease complicating untreated or partially treated Group A Streptococcus (GAS) pharyngitis

PATHOPHYSIOLOGY

Risk factors:
Overcrowding
Poor orodental & personal hygiene limited access to healthcare

Precipitating event:
Group A Streptococcal pharyngitis

Susceptible host:
School going children (5-15 years age)

Repeated group A Streptococcus infections → Priming of immune response

First episode of ARF

Repeated or ongoing infections → RHD

Molecular mimicry between group A Streptococcus antigens and host tissues

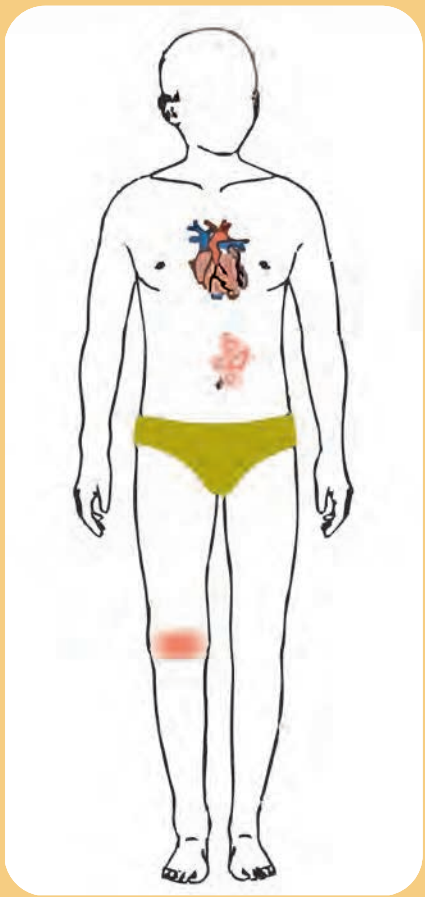
Exaggerated T-cell mediated immune response

CLINICAL PRESENTATION

Arthritis (80%) – Most common manifestation
- Multiple joints
- Migratory – lasts <1 week in a joint
- Large joints – ankles, knees & wrist
- Exquisite tenderness with redness & swelling
- Prompt response to NSAIDs
- Leaves no deformity

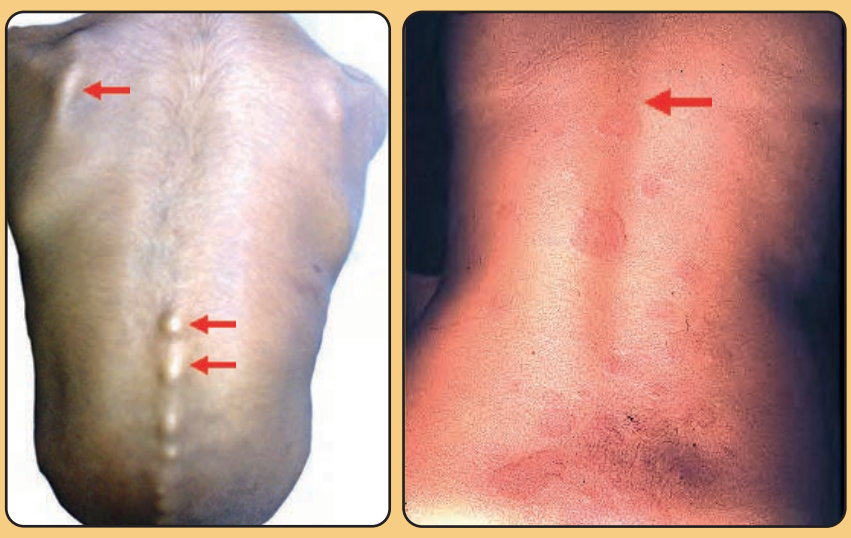
Carditis (50%) – Most devastating manifestation
- Tachycardia
- Dyspnoea
- Heart Failure
- Murmur on auscultation

Chorea (10%) - 2-6 months after streptococcal sore throat
- Quasipurposive, involuntary movements with emotional lability
- Best seen in hands, arms, tongue and face
- Affects fine motor movement like handwriting



Subcutaneous nodules – rare
Painless, pea-sized, hard nodules
On extensor surfaces of limbs, skull and back

Erythema marginatum (5%)
Transient pink macule with fading centre
Mostly located on the trunk and limbs



DIAGNOSIS BASED ON JONES CRITERIA

For all patient populations with evidence of preceding group A streptococcal infection

Diagnosis:	
Initial ARF	2 major or 1 major plus 2 minor Criteria
Recurrent ARF	2 major or 1 major and 2 minor or 3 minor Criteria
Recurrent ARF in RHD	2 minor (No major criteria needed)
Criteria	
Major	
Low-risk populations[a]	***Moderate and high-risk populations***[a]
Carditis (Clinical and/or subclinical)[b]	Carditis (Clinical and/or subclinical)[b]
Arthritis (Polyarthritis only)	Arthritis (Monoarthritis or polyarthritis or polyarthralgia)[c]
Chorea	Chorea
Erythema marginatum	Erythema marginatum
Subcutaneous nodules	Subcutaneous nodules
Minor	
Polyarthralgia[c]	Monoarthralgia
Fever (≥38.5°C)	Fever (≥38°C)
ESR >60 mm/h and/or CRP ≥3 mg/dL	ESR >30 mm/h and/or CRP ≥3 mg/dL
Prolonged PR on ECG (for age) (unless carditis is a major criterion)	Prolonged PR on ECG (for age) (unless carditis is a major criterion)
Essential	
Throat culture or antigen positive for streptococcal sore throat OR elevated ASO titers (>320 U)	

[a]Low-risk populations ARF incidence ≤2/ per 100 000 school-aged children or all-age RHD prevalence of ≤1/per 1000 population per year

[b]Subclinical carditis is pathological echocardiographic valvulitis

[c]Polyarthralgia should only be considered as a major manifestation in moderate-to high-risk populations after exclusion of other conditions. Joint manifestations can only be considered in either the major or minor categories but not both in the same patient

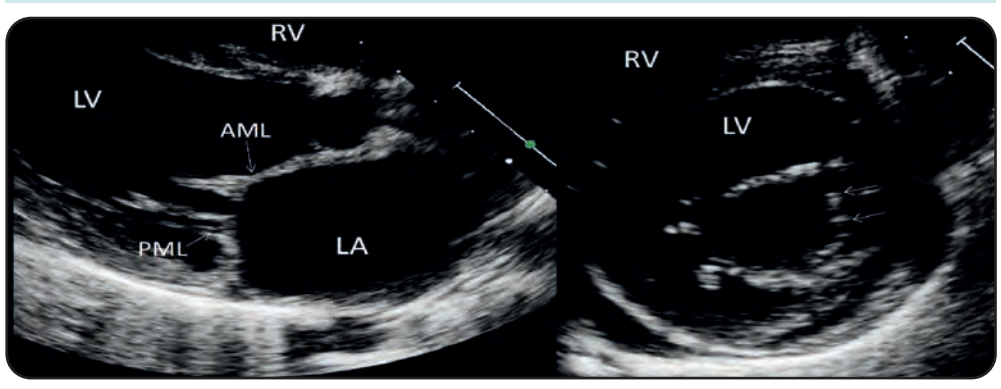
Erythema marginatum and subcutaneous nodules are 'stand-alone major criteria

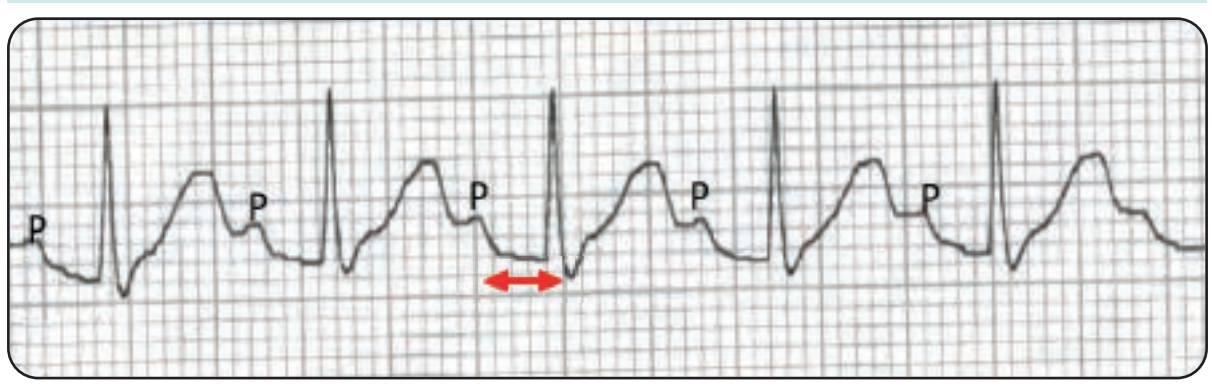
LABORATORY INVESTIGATIONS

Essential		*Optional*
• TLC, DLC	• Chest X ray	• Throat swab antigen
• ESR, CRP	• Anti-streptolysin O	• Throat swab culture
• ECG (12 lead)	• Echocardiogram	• Anti DNAse-B

DIFFERENTIAL DIAGNOSIS

1. Pediatric autoimmune neuropsychiatric disorders (PANDAS)- autoimmune disorder
2. Post streptococcal reactive arthritis (PSRA)- small joint arthritis, poor response to NSAIDs
3. Juvenile rheumatoid arthritis
4. Infective endocarditis





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Standard Treatment Workflow (STW)

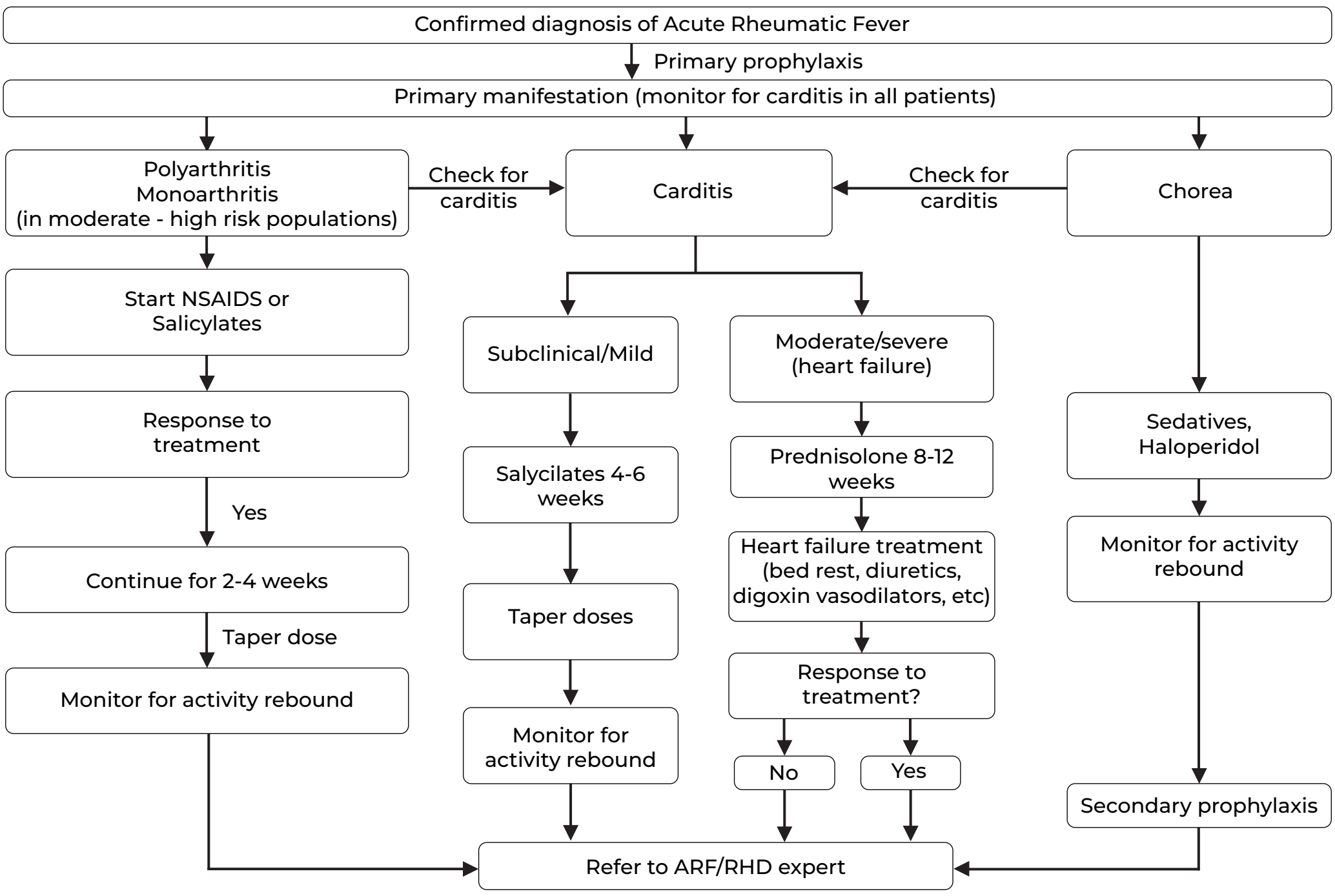
ACUTE RHEUMATIC FEVER

(Continued)

MANAGEMENT

Primary prophylaxis (to Eradicate streptococcus)		
Agent	**Dose**	**Duration**
Benzathine penicillin (Penicillin G)	≤27kg 6,00,000U >27kg 12,00,000U	Once
or		
Phenoxymethyl penicillin(Penicillin V)	≤27kg 250mg/dose <27kg 500mg/dose	10 days
For individuals allergic to penicillin		
Amoxicillin Erythromycin	25-50mg/kg/day divided into 3 doses (maximum 1g/day) 20-40mg/kg/day divided into 2-4 doses (maximum 1g/day)	10 days

Anti-inflammatory therapy & supportive care



Clinical Manifestation	Treatment Schedule	Duration
Moderate/Severe carditis	Prednisolone 2mg/kg/day once daily (Aspirin while tapering Prednisolone)	8-12 WKS
Mild carditis	Aspirin 75-100mg/day divided into 4 doses	2-4 WKS
Polyarthritis	Aspirin 75-100mg/day divided into 4 doses or Naproxen 10-20mg/kg/day	2-4 WKS
Chorea	Carbamazepine 4-10mg/kg/day or Valproic acid 20-30mg/kg/day or Haloperidol 2-6mg/day	Variable depending upon the need of the patient

Secondary prophylaxis

Category of Patient	Duration
Patients without carditis	5 years after the last ARF episode or until 21 years age (whichever is longer)
Patients with carditis but no RHD	10 years after the last acute episode or until 25 years age (whichever is longer)
Patients with RHD who have undergone valve surgery (repair or replacement)	At least until 40 years age (preferably lifelong)

Agent	Dose	Route
Benzathine penicillin (Penicillin G)	≤27kg 6,00,000U >27kg 12,00,000U	Intramuscular
or		
Phenoxymethyl penicillin(penicillin V)	250mg twice daily	Oral
For individuals allergic to penicillin		
Erythromycin	250mg twice daily	Oral

ABBREVATIONS

ARF: Acute Rheumatic Fever
ASO: Antistreptolysin O
CRP: C-reactive protein
DLC: Differential Leukocyte Count
ECG: Electrocardiogram
ESR: Erythrocyte Sedimentation Rate
NSAIDs: Non-Steroidal Anti-Inflammatory Drugs
PANDAS: Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections
RHD: Rheumatic Heart Disease
TLC: Total Leukocyte Count

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☛ INJECTABLE PENICILLIN IS SAFE; ALLERGY IS UNCOMMON

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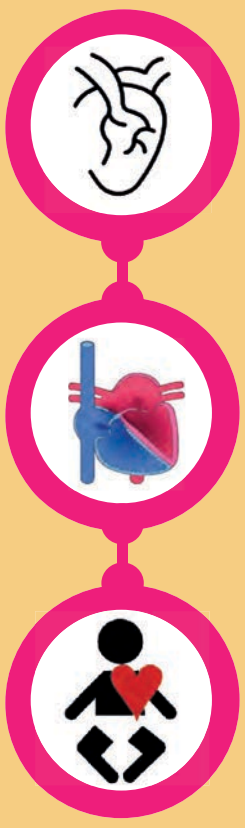
Standard Treatment Workflow (STW)

CRITICAL HEART DISEASE IN THE NEWBORN

ICD-10-P09.5



UNIVERSAL PULSE OXIMETRY SCREENING IS RECOMMENDED (REF 2)

Measurement 1
Pulse oximetry on right arm and 1 foot around 24 hours of age (or earlier, if being discharged)

Fail
Pulse oximetry of 89% or less in either the right arm or foot
Action: Do not repeat pulse oximetry screening, refer for immediate assessment

Retest
a) Pulse oximetry of 90% to 94% in either the right arm or foot or
b) A difference of 4% or more between the right arm and foot
Action: Repeat pulse oximetry measurements

Pass
Pulse oximetry of 95% or more in right arm and foot and a difference of 3% or less between the 2
Action: Do not repeat pulse oximetry screening, provide normal newborn care

Measurement 2
Pulse oximetry on right arm and 1 foot 1 hour after measurement 1

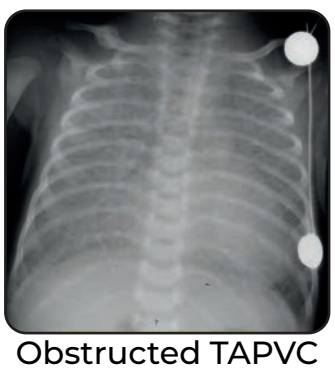
Fail
a) Pulse oximetry of 94% or less in either the right arm or foot or
b) A difference of 4% or more between the right arm and foot
Action: Do not repeat pulse oximetry screening, refer for immediate assessment

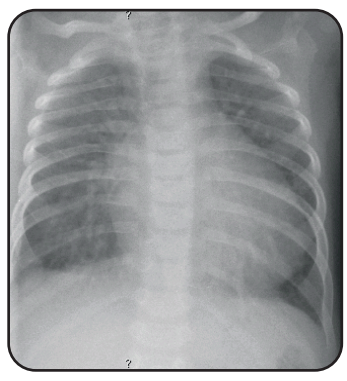
Pass
Pulse oximetry of 95% or more in right arm and foot and a difference of 3% or less between the 2
Action: Do not repeat pulse oximetry screening, provide normal newborn care

Onset of cyanosis	Possible CHD
1st week (Day 1 to 7)	dTGA with intact ventricular septum Hypoplastic left heart or right heart Tricuspid atresia/critical stenosis of PV, MV, AV TOF (severe) or pulmonary atresia TAPVC Truncus arteriosus Ebstein's anomaly
7 days to 1 month	Hypoplastic left heart dextro-Transposition of the Great Arteries (dTGA) TOF Severe PS Truncus arteriosus
Late onset cyanosis	TOF Double outlet right ventricle (DORV) with VSD - PS, dTGA with VSD -PS, Tricuspid atresia with VSD -PS

Hemodynamic approach to CHDs	
Hypotension/shock	Duct dependent systemic circulation (Critical AS, HLHS Severe Interrupted aortic arch) Ventricular dysfunction Arrhythmia with hemodynamic compromise
Severe desaturation	Decreased pulmonary blood flow (duct dependent pulmonary circulation): Pulmonary Atresia, Critical PS TOF with severe PS Ebstein's anomaly Increased PBF & high PA pressure: Transposition
Heart failure	Pulmonary plethora: L -> R shunt With cyanosis/desaturation - CCHD with increased pulmonary blood flow (PBF) With severe desaturation and pulmonary venous hypertension: Obstructed TAPVC

ASK/LOOK/FEEL	CATEGORY	INTERPRETATION
Does the baby have decreased activity and feeds poorly?	Activity and feeding	Decreased activity is a common presentation of heart failure/shock in neonates
Is the baby cyanotic? Pulse Oximetry screen	Cyanosis/Desaturation	Look for bluish discolouration of fingers and tongue. If extremities are blue, to rule out peripheral cyanosis- warm the baby and re check
Is there any evident respiratory distress or Tachypnoea?	Respiration	Chest indrawing/grunting/use of accessory muscles/RR more than 60 per minute
Does the baby have Inappropriate Tachycardia/Bradycardia	Heart Rate	Normal awake new born 100-180 normal sleeping new born 80-160
Is the baby in shock? peripheral temperature	Perfusion	Peripheries cold and clammy OR Cardiac resynchronization therapy(CRT) > 3 seconds, core - difference more than 2 degrees even after warming/external temperature is controlled/ appropriate correction of ambient temperature is done
Is the baby in heart failure?	Heart Failure	Look for Tachypnoea, Tachycardia, Tender Hepatomegaly
Is the baby sucking from the breast normally?	Feeding	Normal: sucking vigorously, no suck rest suck breast cycle, no breathlessness/ forehead sweating while feeding, no prolonged feeding times


Obstructed TAPVC


TGA

APPROACH TO SHOCK

Start PG E1 and refer if:

- Identifiable that femoral pulses are distinctly feeble compared to upper body
- Right arm - Any foot SpO_2 difference more than 3%

Shock
↓
Intravenous/Intraosseous access and fluid resuscitation 10 ml per kg of isotonic fluid, (max 40 ml per kg until perfusion improves or hepatomegaly develops).
Manage Hypothermia, Hypoglycemia, hypocalcemia
Appropriate antibiotic
Monitoring to assess response
↓
Manage shock as per Neonatal shock guidelines
↓
Low threshold for Paediatric Cardiology Evaluation
↓
Refractory Shock, Unlikely to be sepsis - Urgent referral to Pediatric Cardiologist

Septic shock likely if:

- Predisposing maternal and neonatal factors
- Core peripheral temperature difference > 3
- Sepsis Screen Positive

APPROACH TO CYANOSIS

Cyanosis ($<SpO_2<95\%$)
↓
Rule out TAPVC
↓
Chest X-ray S/O whiteout lung/ Ground glass appearance /pulmonary venous hypertension
↓ Yes
If clinical setting makes it unlikely to be Respiratory distress syndrome, Likely obstructed TAPVC -Immediate referral

Significant congenital heart disease likely →

SpO_2 < 80%?
No → Early Paediatric Cardiology Consultation & Echo
Late discharge once diagnosis confirmed
Close SpO_2 monitoring through first week of life (for decrease in SpO_2 on ductal closure)
Heart Failure Management (if only clinical features of CHF) Iron supplementation

Yes → Start PGE1 infusion refer for urgent paediatric cardiology evaluation (within hours)
↑
$SpO_2<80\%$ On serial monitoring

Duct dependent pulmonary circulation (All forms of Pulmonary Atresia/Critical PS) TGA with intact septum

ABBREVATIONS

AS: Aortic Stenosis
AV: Aortic Valve
CCHD: Cyanotic Congenital Heart Disease
CHD: Congenital Heart Disease
HLHS: Hypoplastic Left Heart Syndrome
L->R: Left to Right
MV: Mitral Valve
PA: Pulmonary Artery
PG E1: Prostaglandin E1
PS: Pulmonary Stenosis
PV: Pulmonary Valve
TAPVC: Total anomalous pulmonary Venous Connection
TGA: Transposition of Great Arteries
TOF: Tetralogy of Fallot
TV: Tricuspid Valve
VSD: Ventricular Septal Defect

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INVOLVE A PAEDIATRIC CARDIOLOGIST AS SOON AS CRITICAL CHD IS SUSPECTED

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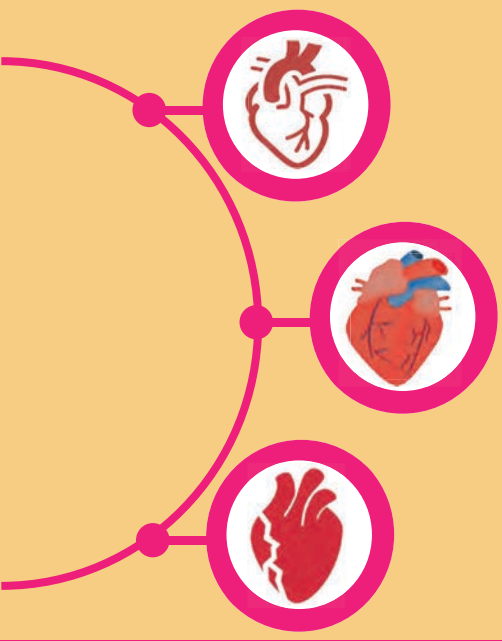
Standard Treatment Workflow (STW)

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

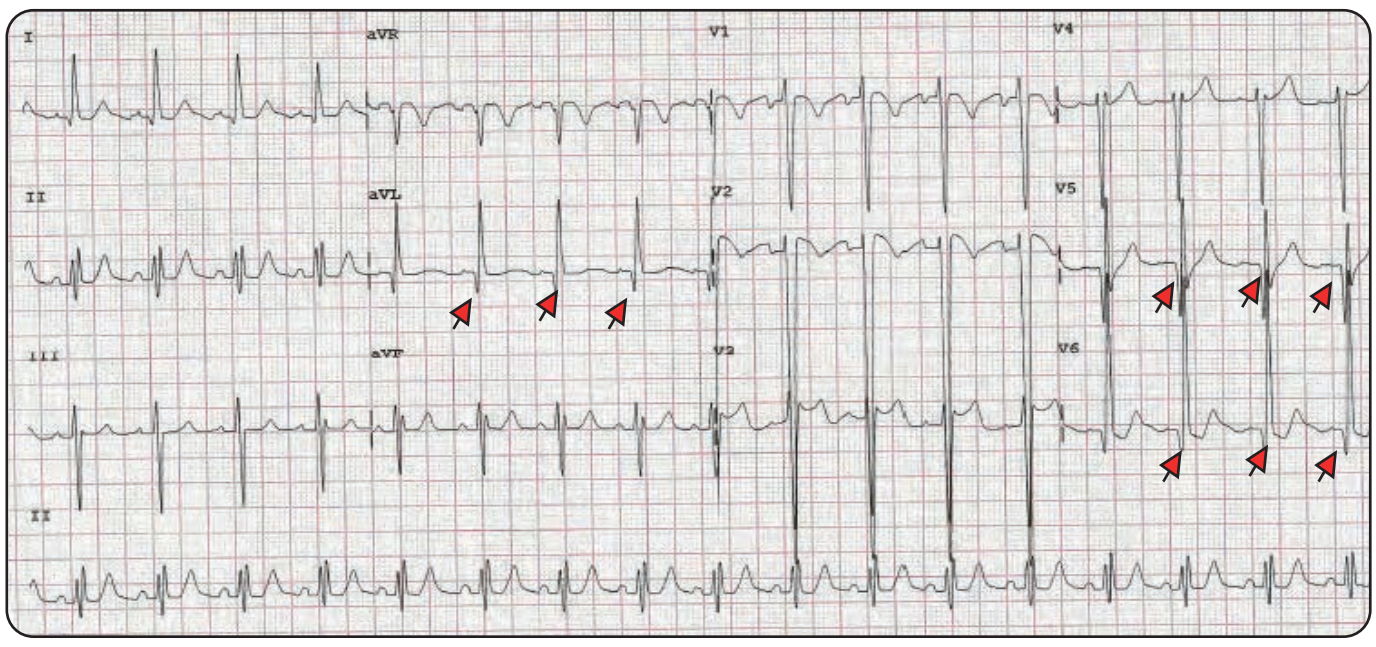
Information on cardiac silhouette, pulmonary vasculature, pulmonary artery dilatation and associated skeletal abnormalities

ECG

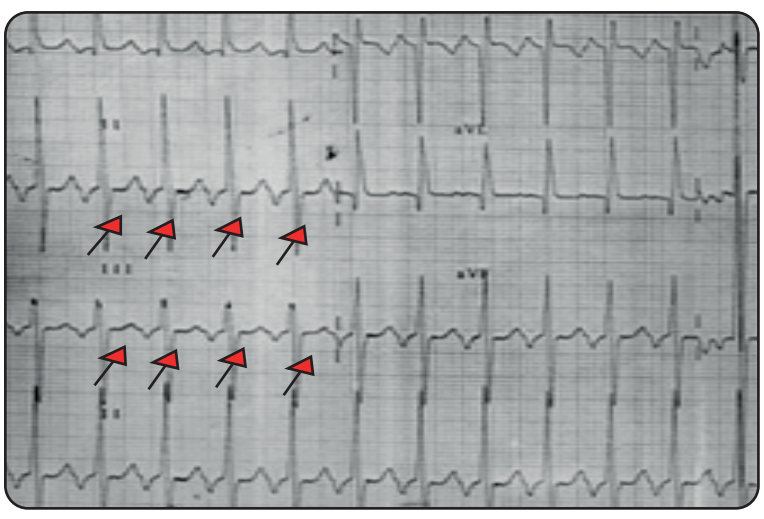
Diagnosis of treatable causes of heart failure such as persistent tachyarrhythmia, ALCAPA and, hypocalcemia. Other specific causes such as Pompe's disease, specific forms of cardiac muscle involvement in muscular dystrophy have ECG manifestations

Echocardiogram

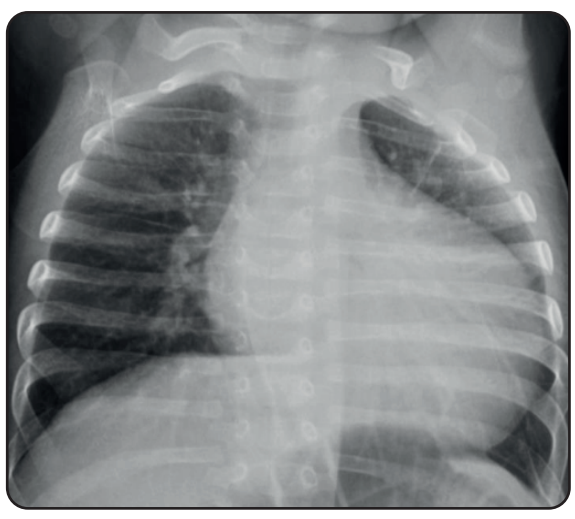
Critically important to accurate diagnosis and tailoring response to therapy



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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Standard Treatment Workflow (STW)

PEDIATRIC HEART FAILURE

(Continued)

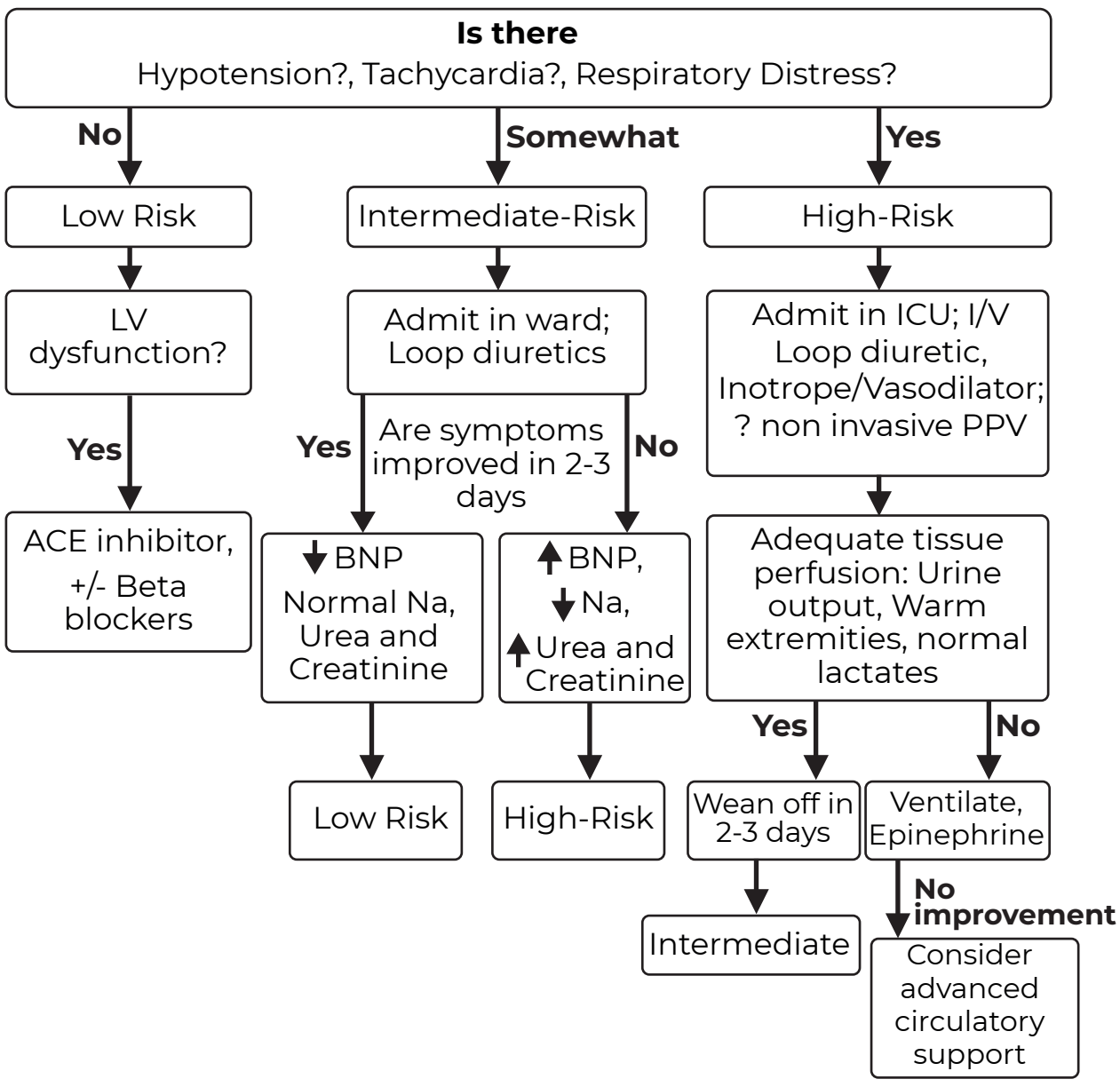
Essential blood tests to be performed in all	Utility
Complete blood count; CRP	Identifying Sepsis, Anemia
Electrolytes and urea, creatinine	Elevated urea, creatinine may indicate decompensated HF or may result from medication side effects. Electrolyte imbalance is a common association of HF and diuretic 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

July/ 2024

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

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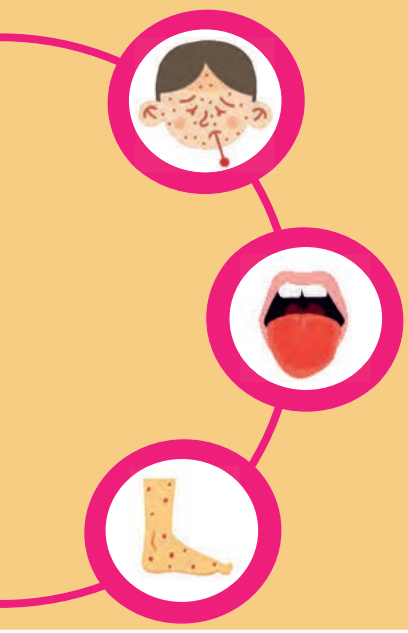
Standard Treatment Workflow (STW)

KAWASAKI DISEASE

ICD-10-M30.3

Any child with fever for more than 5 days should be evaluated for KD



SPECIFIC SIGNS

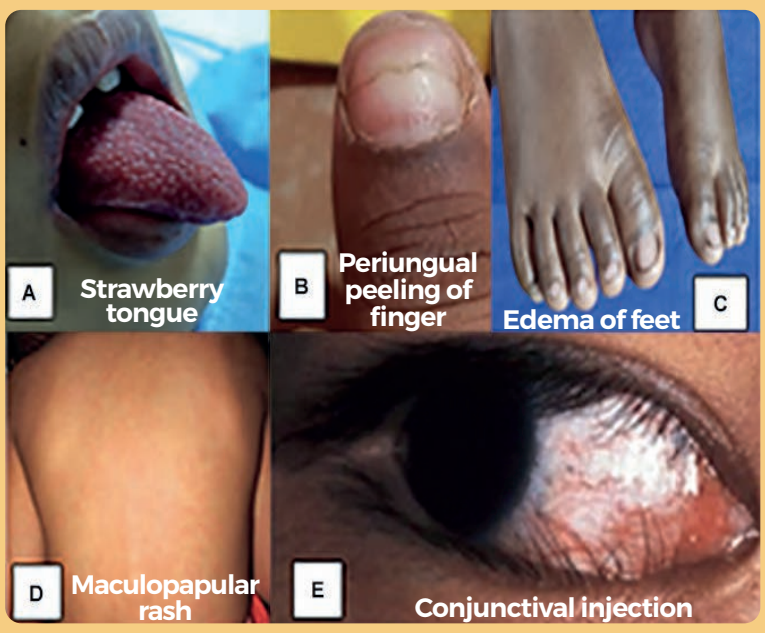
A. Lips and oral cavity:
Erythema & Lip cracking
Strawberry tongue and/or Diffuse erythema of oral & pharyngeal mucosa

B. Changes in extremities:
Erythema of palms, soles
Dorsal edema of hands & feet
Periungual peeling of fingers, toes (2nd & 3rd week)

C. Polymorphous exanthem:
Rash – maculopapular or erythema multiforme-like

D. Eyes: Bilateral conjunctival injection without exudate

E. Cervical lymphadenopathy (>1.5cm diameter) - mostly unilateral



OTHER CLINICAL FINDINGS

GIT: Diarrhea, vomiting, abdominal pain

RS: Cough, rhinorrhea

CVS: Signs of CCF, new onset murmur, gallop

MUSCULOSKELETAL: Arthritis, arthralgia

CNS: Extreme Irritability

OTHERS: Induration at BCG scar site

EXCLUDE OTHER COMMON DISEASES WITH SIMILAR FINDINGS

INFECTIONS
Viral: Measles, Adenovirus, Enterovirus, EBV, CMV
Bacterial: Scarlet fever, Bacterial cervical lymphadenitis, Meningococcemia, leptospirosis

RHEUMATOLOGICAL DISEASE
Systemic onset juvenile idiopathic arthritis

OTHERS
TSS, SSSS, Drug hypersensitivity reaction, SJS

CONSIDER KD IN DIFFERENTIAL DIAGNOSIS IF PROLONGED FEVER OCCURS IN:

- Infants < 6 months with irritability
- Infants with unexplained aseptic meningitis
- Infants or children with unexplained or culture –negative shock
- Infants or children with cervical lymphadenitis unresponsive to antibiotics
- Infants or children with retropharyngeal phlegmon unresponsive to antibiotics

WHEN NOT TO CONSIDER KD?

- Bullous/vesicular rash
- Exudative conjunctivitis
- Exudative pharyngitis
- Ulcerative oral lesions
- Generalized lymphadenopathy
- Splenomegaly

≥ 4 signs: **CLASSICAL KD** → Investigations → Treatment

Fever (≥ 5 days) & < 4 signs: OR Infants with fever (≥ 7 days) → Consider **INCOMPLETE KD** → Refer to a District Hospital/ Tertiary Pediatric Hospital → Reassess Patient characteristics

Investigations:
- CRP < 3 mg/dL and ESR< 40 mm/hr → Serial clinical and lab re-evaluation if fever persists; ECHO if peeling develops under nailbeds
- CRP≥ 3 mg/dL and/ or ESR≥ 40 mm/hr → ≥ 3 lab findings:
  - Anemia for age
  - Platelet count ≥ 4.5 lakhs (> 7 days of fever)
  - Albumin ≤ 3 g/dL
  - Elevated SGPT
  - WBC ≥ 15,000/mm³
  - Urine > 10 WBC/hpf

  or
  - Positive Echocardiogram

  → Yes → **INCOMPLETE KD** → Treatment

  → No → Serial clinical and lab re-evaluation if fever persists; ECHO if peeling develops under nailbeds

DISCUSS WITH RHEUMATOLOGIST

- Infants with KD
- Children with coronary dilatation at time of diagnosis
- Children with shock and myocarditis
- Children who have features of secondary MAS

May need primary intensification of therapy in addition to IVIG (Infliximab, Steroids, Cyclosporine, etc.)

MANAGEMENT

INVESTIGATIONS

- CBC
- CRP
- ESR
- Serum electrolytes
- LFT
- Urine microscopy
- Echocardio-gram

WHAT TO LOOK FOR?

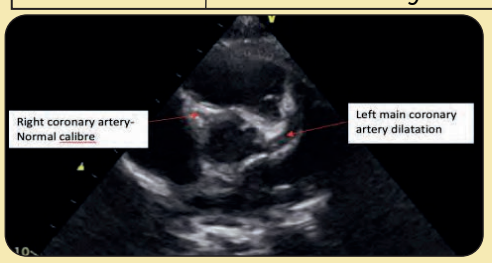
- **CBC:** Leukocytosis –Neutrophilia, Anemia, Thrombocytosis (in 2nd week)
- **CRP-** ↑
- **ESR-** ↑
- **LFT:** SGOT, SGPT - ↑, Albumin ↓
- **Serum electrolytes** – Sodium ↓
- **Urine microscopy-** Sterile pyuria
- **ECHO-** Coronary artery dimensions, perivascular brightness, lack of tapering, LV dysfunction, mitral regurgitation, pericardial effusion

ECHOCARDIOGRAPHY - TO BE DONE BY A PAEDIATRIC CARDIOLOGIST

Positive ECHOCARDIOGRAM:
Any one of the below-
- RCA or LAD Z score: ≥ 2.5
- Coronary artery aneurysm
- ≥ 3 of the following:

LV dysfunction, Mitral regurgitation, pericardial effusion, RCA or LAD Z score: 2 - 2.5

Z-SCORE CLASSIFICATION	
< 2	Normal
2-2.5	Only dilatation
≥ 2.5 to < 5	Small aneurysm
≥ 5 to < 10	Medium aneurysm
≥ 10	Giant aneurysm



2-D ECHO imaging:
Aim for highest resolution & frame rate possible
Phased array transducer with highest frequency possible
- Narrow sector width
- Adjust focus to region of interest
- Reduce depth
- Zooming in
- Optimize gain

TREATMENT

WHEN TO START IVIG?

- In children who meet diagnostic criteria as soon as possible (ideally within 10 days of fever onset)
- Even after 10 days of illness if evidence of systemic inflammation is present (elevated ESR/ CRP) with fever
- Recurrent KD (repeat episode after complete resolution of previous episode)

Unavailability of ECHO should not delay IVIG if diagnostic criteria are met

- Intravenous Immunoglobulin-IVIG (2g/kg) as a single infusion over 10-12 hours
- Aspirin 80-100 mg/kg/day in 4 divided doses –till child is afebrile or 48 to 72 hrs after cessation of fever

→ **Aspirin:** 3-5 mg/kg/day for 6 to 8 weeks

LONG TERM THROMBOPROPHYLAXIS FOR CORONARY ARTERY INVOLVEMENT

CORONARY ARTERY	DRUG	DURATION
No involvement	Aspirin* 3-5 mg/kg/day	6-8 weeks
Only dilatation	Aspirin* 3-5 mg/kg/day	6-8 weeks
Small aneurysm	Aspirin* 3-5 mg/kg/day	Till aneurysm resolves (Consult pediatric cardiologist)
Medium aneurysm	Aspirin* 3-5 mg/kg/day + Clopidogrel 0.2-1mg/kg/day	
Giant aneurysm	Aspirin* 3-5 mg/kg/day + Anticoagulation (Warfarin: 0.2 mg/kg/day loading, then 0.1mg/kg/day or LMWH 1mg/kg/day)	

*If patient is intolerant/resistant to Aspirin - use Clopidogrel

TREATMENT OPTIONS FOR IVIG RESISTANCE (PERSISTENT OR RECRUDESCENT FEVER 36 HOURS AFTER THERAPY WITH IVIG)

DRUGS	DOSE	DURATION
IVIG (second infusion)	2g/kg IV	Single dose
Pulse methyl prednisolone followed by Oral prednisolone in tapering doses	Intravenously (10-30 mg/kg/day) 2mg/kg	3-5 days Till CRP is normal, then taper over 2-3 weeks
Infliximab	5mg/kg IV over 3-4 hours	Single dose

ABBREVATIONS

CBC: Complete Blood Count
CMV: Cytomegalovirus
CRP: C-reactive Protein
EBV: Epstein-Barr Virus
ESR: Erythrocyte Sedimentation Rate
KD: Kawasaki Disease
LAD: Left anterior Descending Artery
LFT: Liver Function Test
LMWH: Low Molecular Weight Heparin
LV: Left Ventricle
MAS: Macrophage Activation Syndrome
RCA: Right coronary Artery
SGOT: Serum Glutamic Oxaloacetic Transaminase
SGPT: Serum Glutamic-Pyruvic Transaminase
SJS: Stevens-Johnson Syndrome
SSSS: Staphylococcal Scalded Skin Syndrome
TSS: Toxic Shock Syndrome
WBC: White Blood Cell

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DELAY IN DIAGNOSING KAWASAKI DISEASE CAN RESULT IN ADVERSE CLINICAL OUTCOMES

July/ 2024

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Standard Treatment Workflow (STW)

LEFT TO RIGHT SHUNT LESIONS

ICD-10-Q21.8

INTRODUCTION

- Most common type of congenital heart defects
- One of the common causes of infant morbidity and mortality
- Majority of the lesions are easily correctable if detected on time

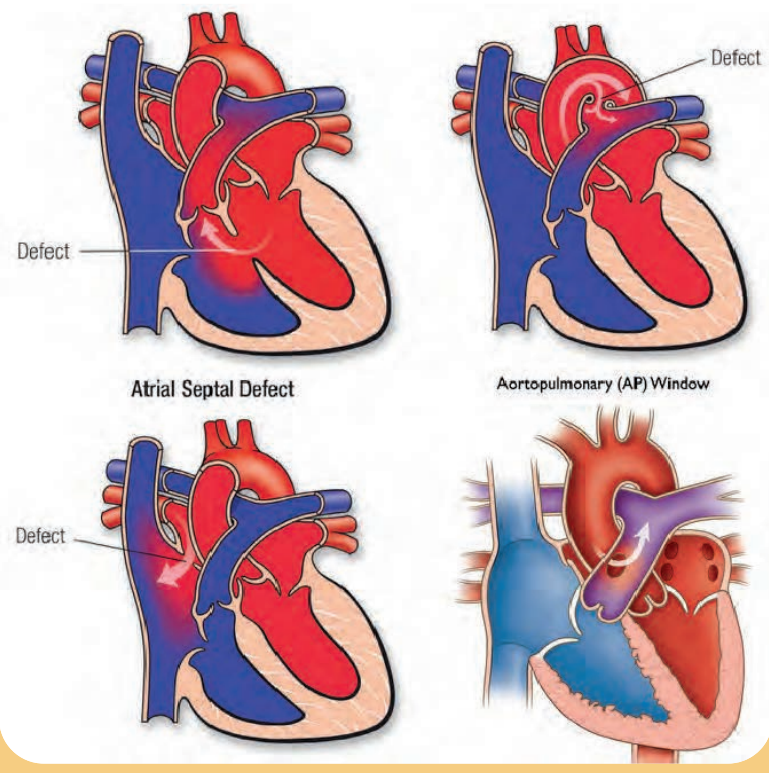
PHYSIOLOGY

- Left to right shunt lesions lead to passage of oxygenated blood from left side of heart to right side and into the lungs
- As a result there is increased flow to the lungs and over circulation of blood within the lungs and left side of the heart
- Majority of symptoms of shunt lesions are due to this over circulation

COMMON LEFT TO RIGHT SHUNT LESIONS

- **Pre-tricuspid shunts:**
  - Atrial septal defect (ASD): Usually asymptomatic. Presents commonly as incidentally detected murmur
- **Post-tricuspid shunts:**
  - Ventricular septal defect (VSD)
  - Patent ductus arteriosus (PDA)
  - Aorto-pulmonary window (APW)

Large post-tricuspid shunts present early (usually by 1.5-2 months of age) with signs of cardiac failure like feeding and breathing difficulty along with failure to thrive



MANAGEMENT

WHEN TO SUSPECT?

1. Failure to thrive (weight less than 3rd centile for age, drop in weight by more than 2 major centile lines)
2. Feeding difficulty (suck-rest-suck cycle) with forehead sweating (cold sweats)
3. Repeated chest infections/one life threatening infection
4. Baseline tachypnea with subcostal and intercostal retractions:
   - Rate > 60/min in less than 1 year old
   - Rate > 50/min between 1-2 year old
5. Tachycardia:
   - Rate > 160/min in less than 1 year old
   - Rate >140/min between 1-2 year old
6. Bounding (high volume) pulse (in PDA and APW)
7. Precordial bulge with active precordium
8. Loud second heart sound, gallop rhythm, ejection systolic murmur, mid-diastolic murmur (Large shunts may not have loud murmurs)
9. Hepatomegaly
10. Dysmorphic features: Down syndrome are known to be associated with Atrioventricular septal defect(AVSD)
11. Abnormal peripheral pulses especially feeble lower limb pulses

Timely referral to higher centre with pediatric cardiac facility

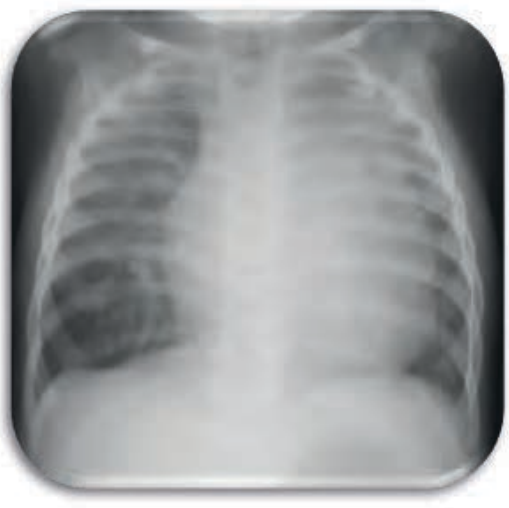
- Shunt lesions are confirmed by echocardiography
- Large post tricuspid shunts require early referral

Drugs

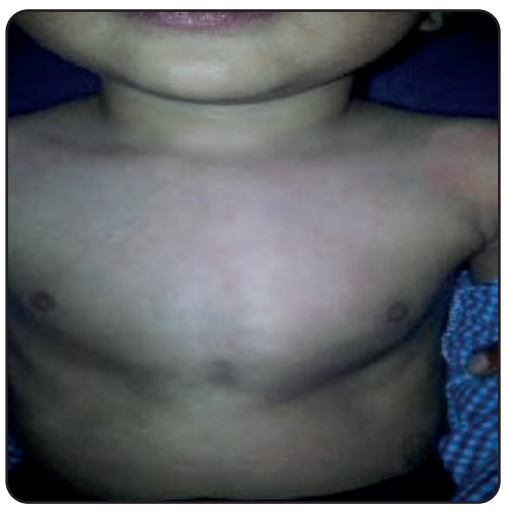
- Furosemide: 1-2 mg/kg/dose twice or thrice daily (reduce or temporarily stop during diarrhea or vomiting). Oral suspension contains 10 mg/ml. So can be given as 0.1 ml/kg/dose twice or thrice daily
- Add Spironolactone if Furosemide is administered more frequently than once daily
- Digoxin: 5 microgram/kg/dose twice daily. Oral preparation contains 50 microgram/ml. So can be given as 0.1 ml/kg/dose twice daily

General Advice

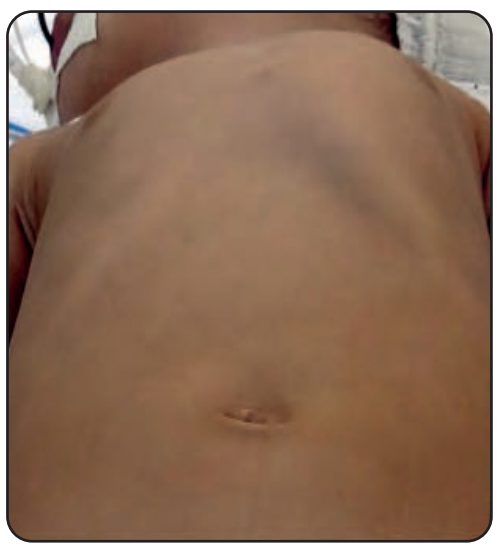
- Educating parents about importance of maintaining hygiene to prevent infections
- Promoting breastfeeding if tolerated. If breastfeeding is difficult then teach gavage/spoon feeding, preferably with expressed breast milk
- Use top milk in case of reduced breastmilk output. Average volume intake should be approximately 120 mL/kg/day
- Include energy dense weaning foods in those beyond 6 months of age
- Continue vaccination as per Indian Academy of Pediatrics (IAP) schedule
- Vitamin D3, calcium and iron supplementation to be continued as per IAP recommendations and clinical requirement



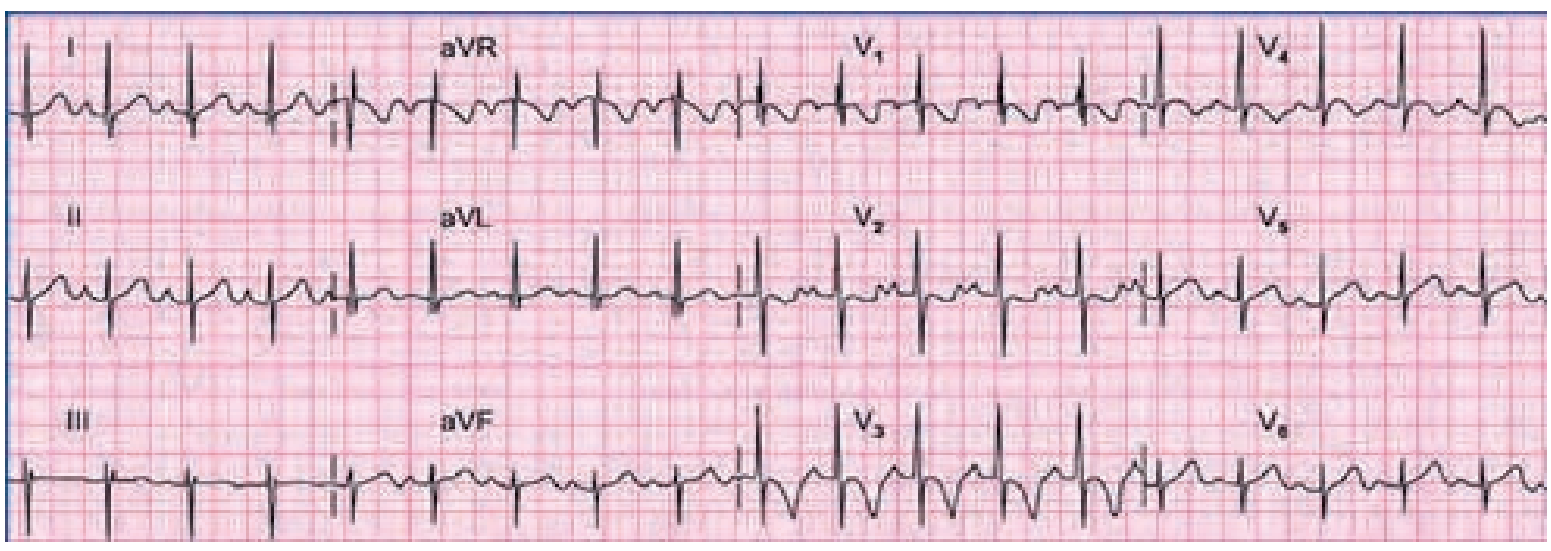
Cardiomegaly & increased vascular markings in shunt lesion



Harrison sulcus



Precordial bulge (left side)



12-lead ECG showing left axis deviation in a patient with AV septal defect

INVESTIGATIONS

Essential

- X-ray Chest, Echo
- ECG - To watch for unexpected abnormal axis, rate, rhythm and QRS complex
- CBC, Electrolytes - Depending on clinical conditions and specific clinical circumstances

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TIMELY CORRECTION OF SHUNT LESION ENABLES NEAR NORMAL QUALITY OF LIFE

July/ 2024

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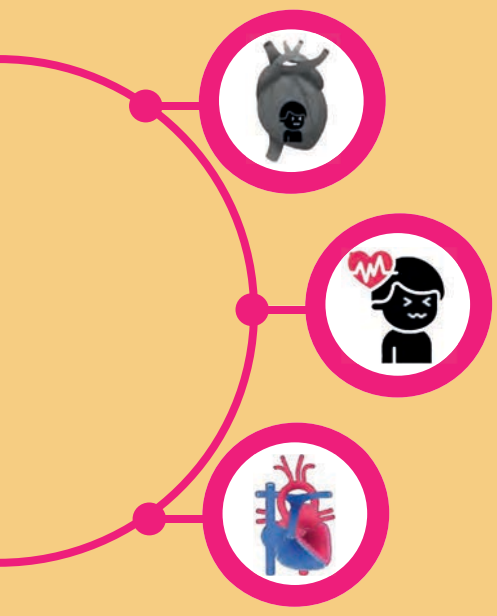
Standard Treatment Workflow (STW)

TACHYARRHYTHMIA

ICD-10-P29.11



SUSPECTING TACHYARRHYTHMIA SYMPTOMS

- Palpitations/chest discomfort
- Parents may report increased precordial activity or observe neck pulsations
- Unexplained lethargy
- Syncope/presyncope: Relatively rare in children but potentially serious

SIGNS

- Tachycardia out of proportion to clinical condition
- Irregular heart rate
- Unexplained heart failure

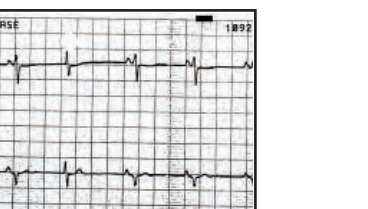
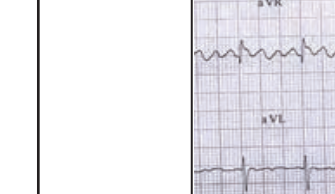
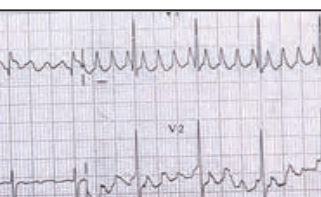
KEY QUESTIONS

- Is there hemodynamic instability?
- Can the heart rate be explained by clinical condition (Fever etc.)
- Is the arrhythmia incessant or episodic?
- Is there an underlying structural heart disease?
- Is this a re-entrant arrhythmia or does it involve an automatic focus?

MANAGEMENT

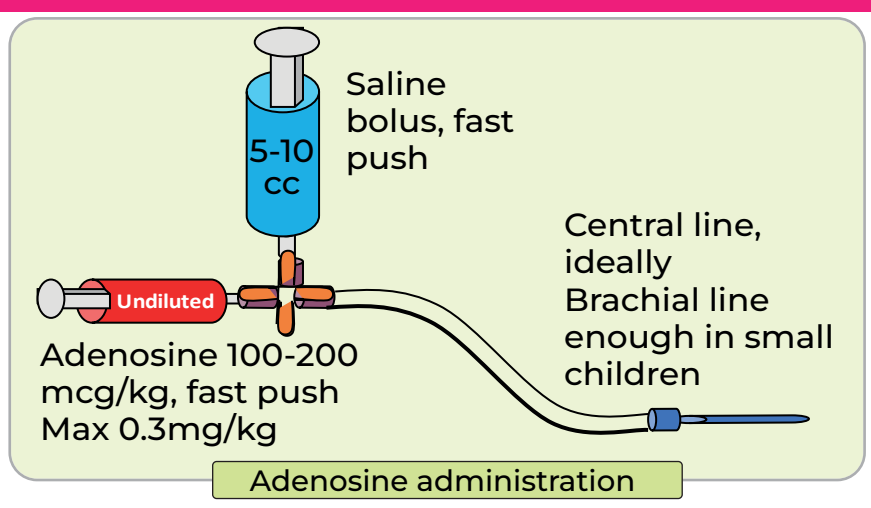
Hemodynamic Stability

Stable	Unstable
Common Stable/minimally distressed Good perfusion	Uncommon Distressed In shock Poor perfusion; pulse not felt
	Connect to Defibrillator paddles

Narrow QRS tachycardia	Regular Wide QRS tachycardia	Irregular Wide QRS tachycardia
		
Synchronized Cardioversion 1J/Kg		Defibrillation 2 J./Kg

Obtain

1. 12 lead ECG; Limb leads alone if child does not cooperate (If ECG machine is unavailable, a video recording of the monitor must be obtained)
2. Reliable I/V access; Proximal sites preferred



Adenosine administration

REGULAR TACHYCARDIA

Sinus tachycardia suggested by:

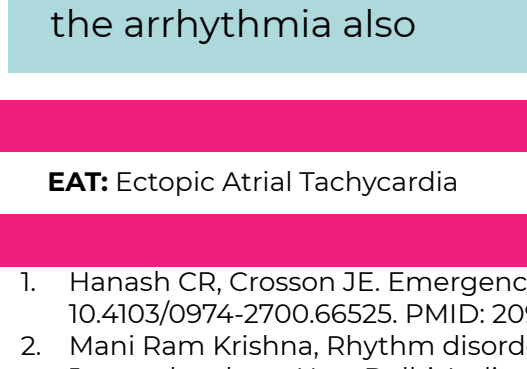
- Heart rates <220 -age
- Subtle variations in rates
- Associated fever/systemic illness other conditions
- Bronchodilators/ Adrenaline nebulization
- Normal p prior to every QRS

Tachyarrhythmia suggested by:

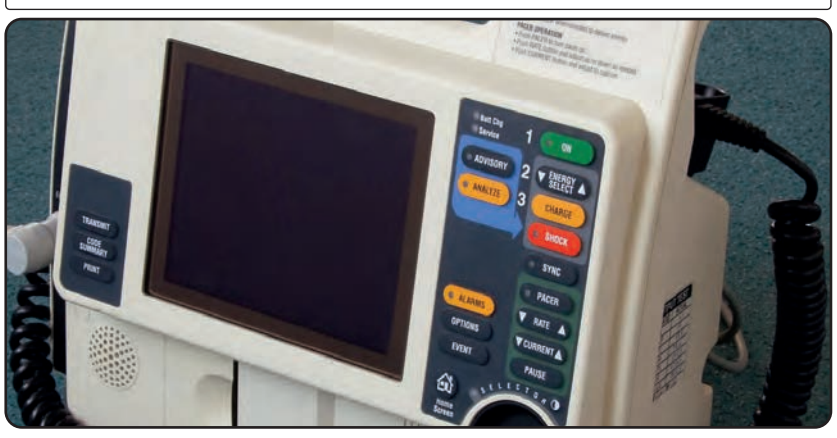
- Fixed rates often >220 -age
- Tachycardia not explained by clinical condition
- Abnormal ECG (p waves not clearly seen or different from sinus rhythm or dissociated)
- Adenosine administration with ECG record is often diagnostic

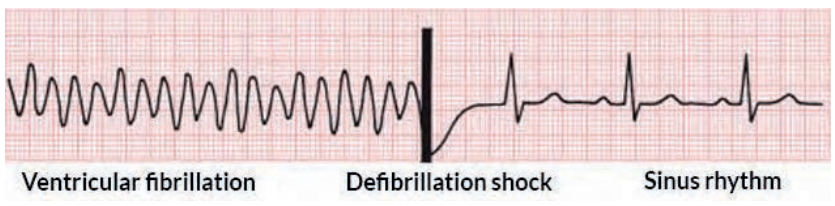
Obtaining ECG during arrhythmia is of great value as it enables precise diagnosis and treatment. All efforts must be made to document the tachyarrhythmia and it's response to treatment

Adenosine

Sudden termination	No effect	Slow and unmask
		
Re-entrant supraventricular tachycardia	Sinus tachycardia, Junctional ectopic tachycardia, EAT	Atrial Flutter, ectopic atrial tachycardia (EAT)

Defibrillation

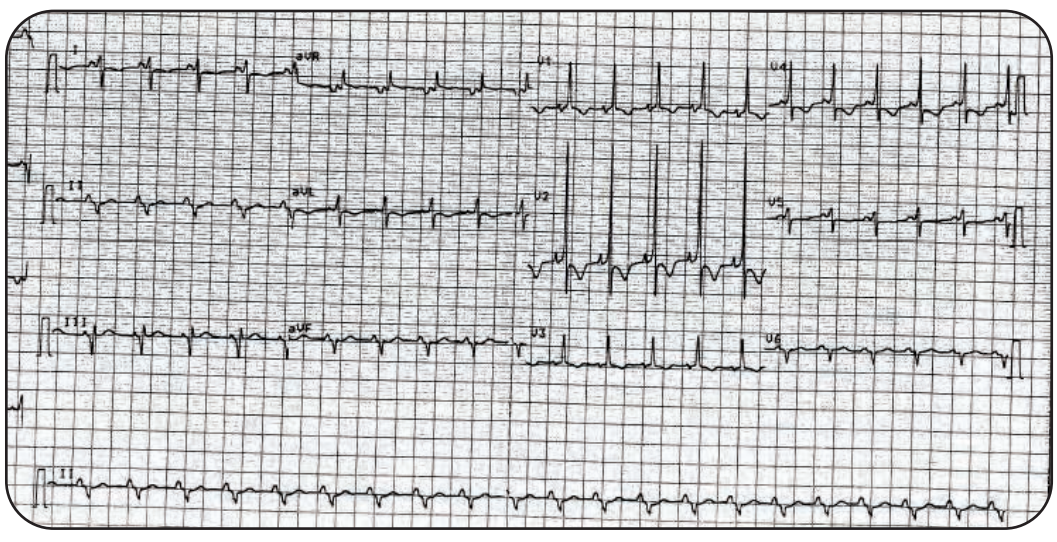




0.5-2 J/Kg synchronized for suspected SVT/VT
2-4 J/Kg for VF; should not be synchronized

Adenosine

- Proximal access
- Connect three-way to I/V port
- Adenosine 100-200 mcg/Kg rapid I/V push followed immediately by 5-10 ml saline bolus
- Always record Electrocardiogram (ECG) during administration
- Always record Electrocardiogram (ECG) after treating the arrhythmia also



12 lead ECG recorded after termination of the tachycardia showing a clear substrate in the form of pre-excitation

ABBREVATIONS

EAT: Ectopic Atrial Tachycardia **SVT:** Supraventricular Tachycardia **VT:** Ventricular Tachycardia

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ALWAYS TRY IDENTIFY AND DOCUMENT THE ARRHYTHMIA PRIOR TO TREATMENT