July/ 2024

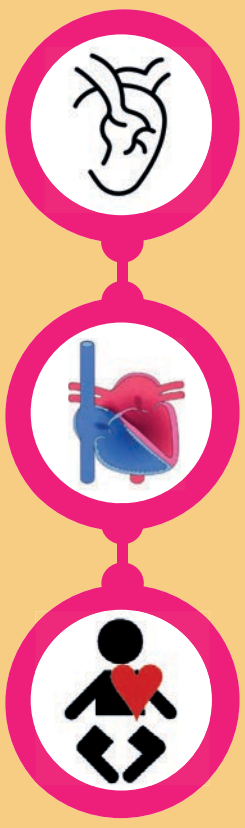
Department of Health Research
Ministry of Health and Family Welfare, Government of India

INDIAN COUNCIL OF MEDICAL RESEARCH
Serving the nation since 1911

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

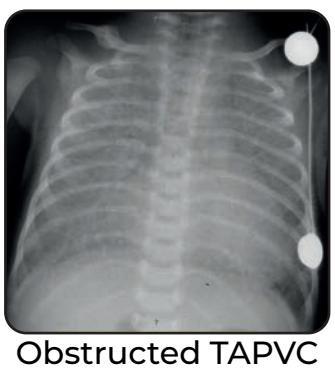
Fail
a) Pulse oximetry of 94% or less in either the right arm or foot or
b) A difference of 4% or more between the right arm and foot
Action: Do not repeat pulse oximetry screening, refer for immediate assessment

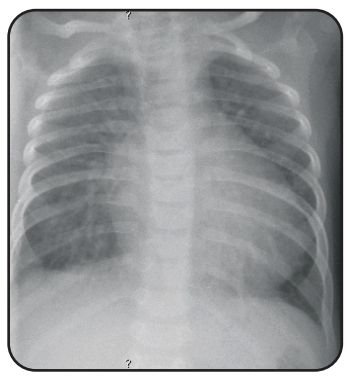
Pass
Pulse oximetry of 95% or more in right arm and foot and a difference of 3% or less between the 2
Action: Do not repeat pulse oximetry screening, provide normal newborn care

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

REFERENCES

1. Gupta SK. Congenital heart disease. In Agarwal R, Deorari A, Paul V, Sankar MJ, Sachdeva A (Eds), AIIMS protocols in Neonatology. Noble Vision Medical Books Publishers, New Delhi 2019. Page 150-164
2. Martin GR, Ewer AK, Gaviglio A, Hom LA, Saarinen A, Sontag M, Burns KM, Kemper AR, Oster ME. Updated Strategies for Pulse Oximetry Screening for Critical Congenital Heart Disease. Pediatrics. 2020 Jul;146(1):e20191650. doi: 10.1542/peds.2019-1650. Epub 2020 Jun 4. PMID: 32499387

INVOLVE A PAEDIATRIC CARDIOLOGIST AS SOON AS CRITICAL CHD IS SUSPECTED