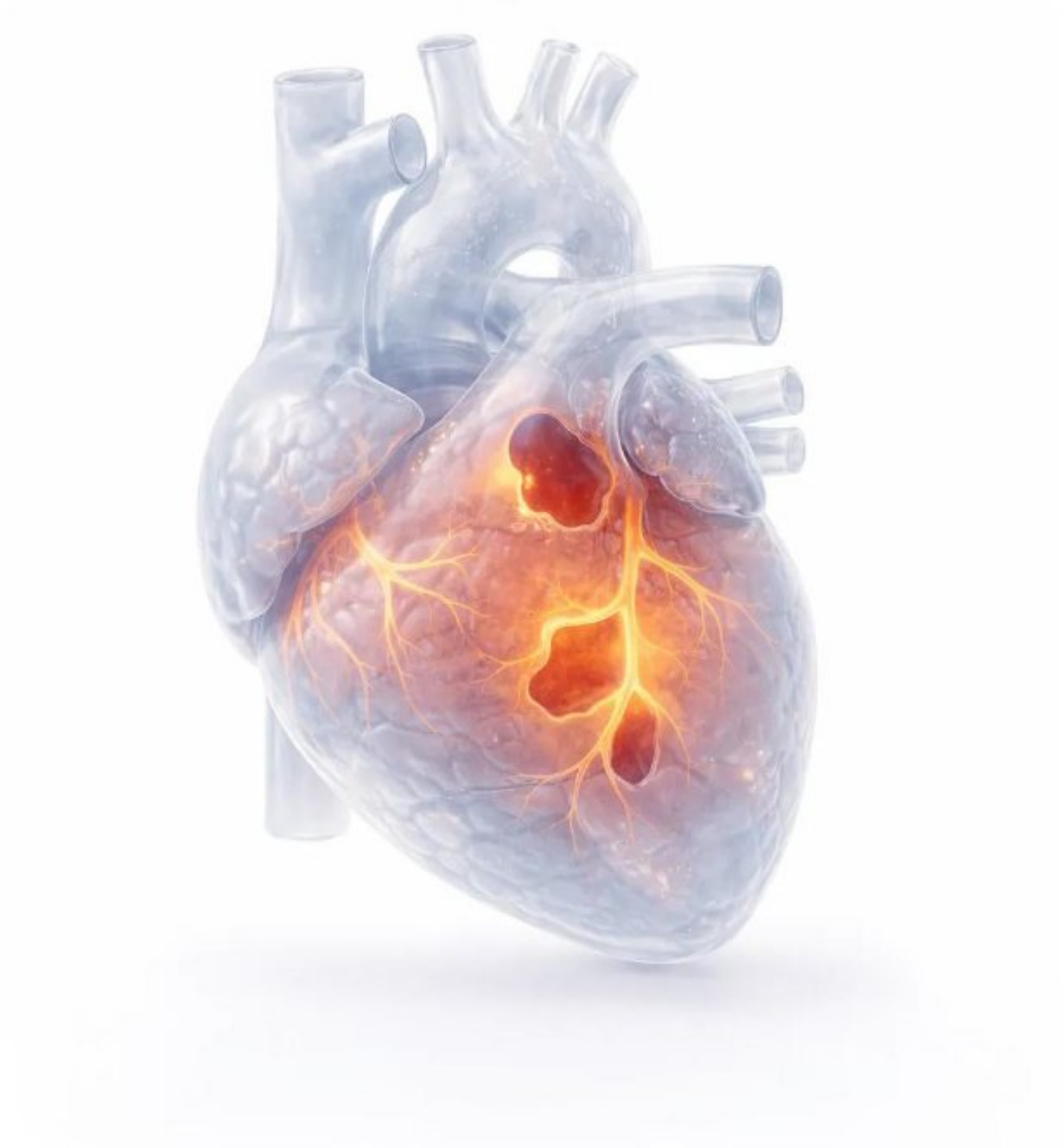
CONGENITAL HEART DISEASES

National Standard Treatment Guideline



Ministry of Health
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ADB

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National Standard Treatment Guidelines

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GUIDELINES DEVELOPMENT METHODOLOGY

The development of the Maldives Standard Treatment Guidelines (STGs) followed a structured, evidence-informed, and consensus-driven methodology adapted from internationally accepted guideline-development standards and the Delhi Society for Promotion of Rational Use of Drugs (DSPRUD) model. The process combined systematic evidence retrieval, critical appraisal, contextual adaptation, and multidisciplinary expert review to ensure feasibility, clinical relevance, and national ownership.

1. Determining Scope and Priority Conditions

Priority clinical conditions were identified through consultation with national programme managers, specialty clinicians, and health-system stakeholders. Selection criteria included: (i) major causes of morbidity and mortality, (ii) observed variation in clinical practice or prescribing patterns, (iii) potential to improve patient outcomes, and (iv) the feasibility of implementation across health-facility levels in Maldives. The final list of diseases reflected national epidemiology, service-delivery capacity, and essential-medicine availability.

2. Identification of Existing Evidence and Source Guidelines

A targeted search strategy was used to identify high-quality existing clinical guidelines. Searches were conducted across international guideline repositories (e.g., WHO, NICE, SIGN and other intergovernmental bodies, international and national guideline repositories, specialty societies and professional associations.

3. Quality Appraisal of Source Guidelines

Retrieved guidelines were screened for transparency of development, methodological rigour, clarity of recommendations, applicability to health-system reality, editorial independence. Guidelines were included if they met the Institute of Medicine (IOM) definition of a clinical guideline and addressed treatment or management of priority conditions. Guidelines that did not meet minimum quality standards, review articles, diagnostic criteria, or technical standards were excluded.

4. Adoption, Adaptation, and Contextualization

The guideline-development team employed an adopt–adapt–contextualize model:

- **Adoption**: High-quality recommendations that aligned with Maldivian health-system realities were retained without modification.
- **Adaptation**: Recommendations were modified when local considerations such as diagnostic capacity, medicine availability, workforce skills, referral pathways, or cost constraints affected feasibility.

- **Contextualization**: Where evidence was absent or inconclusive, conditional recommendations were formulated based on expert consensus, with explicit consideration of pragmatism, safety, and local workflows.
  Medicines were selected in alignment with the Maldives National Essential Medicines List (NEML), based on suitability, efficacy, safety, and availability.

5. Expert Consensus and Multidisciplinary Input

Draft recommendations were initially prepared by experts from the DSPRUD, India, providing a strong methodological foundation for the process. Building on this, a collaborative and participatory process brought together clinicians from internal medicine, paediatrics, obstetrics-gynaecology, surgery, emergency medicine, endocrinology, cardiology, general practitioners, and public health representing different levels of healthcare. Consensus was achieved through moderated discussions, iterative revisions, and resolution of divergent views. For topics lacking strong evidence, recommendations were derived from expert clinical judgment grounded in extensive practice experience.

6. Drafting, Peer Review, and Validation

Each guideline section was organized in a standard format including key clinical features, essential investigations, non-pharmacological management, pharmacological therapy (with step-up/step-down options where relevant), referral criteria, paediatric considerations, and follow-up requirements. Drafts were peer-reviewed by senior clinicians and national experts. Reviewer comments were systematically integrated to strengthen clarity, accuracy, and applicability.

7. Addressing Conflicts of Interest

All contributors declared the absence of conflicts of interest. Individuals with potential or perceived conflicts were excluded from authorship or decision-making roles.

8. Updating and Future Revisions

The STGs were conceptualized as a living document. Future updates will incorporate new scientific evidence, changes in essential-medicine availability, national programme priorities, and user feedback from clinicians. Periodic review cycles will ensure the continued relevance and reliability of recommendations.

9. Distinctive Features of the Guidelines

Developed through a collaborative process involving a large group of multidisciplinary experts from different levels of healthcare, the guidelines incorporate the following distinctive features:

- **Diagnostic Assumption and Confirmation**: While assuming that an initial diagnosis has been established by the healthcare provider, the guidelines provide essential information for confirming diagnoses. This includes a comprehensive overview of major signs and symptoms, descriptions of confirmatory tests, and clear guidance on practices that are prohibited, discouraged, or unreliable—promoting evidence-based medicine supported by relevant references.
- **Comprehensive Treatment Approach**: The guidelines offer a systematic, up-to-date framework for managing medical conditions across the continuum of care. They begin at the primary care level and extend to secondary and tertiary care, incorporating protocols for treatment response assessment and referral criteria as integral components.
- **Diverse Treatment Modalities**: Recommendations encompass both non-pharmacological and pharmacological interventions and surgical intervention where applicable, providing flexibility for individualized treatment plans. Cautionary notes are included where necessary to ensure safe and effective use of therapies.
- **Assessment and Referral Criteria**: Clear criteria and goals for evaluating patient response to treatment are provided, along with guidance on when referral to higher levels of care is warranted ensuring continuity and comprehensiveness in patient management.

ACKNOWLEDGEMENTS

The Government of the Republic of Maldives is committed to ensuring universal access to quality health services for all citizens. The Constitution of Maldives mandates the progressive realization of rights, including the right to good standards of health care for the population. In line with this national commitment, standardized quality health services are regarded as the foundation of a strong and equitable healthcare system.

This important work would not have been possible without the cooperation and support of many individuals and institutions. We express our sincere appreciation to the Honourable Minister of Health, Abdullah Nazim Ibrahim, for his leadership, commitment, and continuous guidance throughout the development process. We are grateful to WHO and ADB for their significant contribution, support, and technical assistance.

Our heartfelt gratitude is extended to the technical lead and editor, Dr. Sangeeta Sharma, Professor, Neuropsychopharmacology, IHBAS and President, Delhi Society for Promotion of Rational Use of Drugs (DSPRUD), and her team. We express our deepest appreciation to the Maldivian and DSPRUD experts and contributors who played a pivotal role in this process. Their technical expertise and dedication to adapt the standards to the Maldivian context have been instrumental in the development and finalization of these guidelines. The time, experience, generous sharing of knowledge and insights contributed by all parties have not only enriched the work but also have been invaluable in making these standards practical, locally acceptable, and aligned with the needs of the resident population.

It is important to acknowledge the immense efforts, involvement, timely coordination, collaboration, and dedication of the Quality Assurance and Regulation Division team who made it possible for these Clinical Treatment Guidelines to come into existence.

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CONGENITAL HEART DISEASES

QUICK REFERENCE GUIDE

SECTION I

SCREENING & MANAGEMENT OF CRITICAL CONGENITAL HEART DISEASE

CHD encompasses a wide range of structural cardiac abnormalities present from birth, resulting from disruptions in normal fetal heart development. A subset, i.e., critical CHD can cause hypoxemia or shock soon after delivery. Many babies look well initially and deteriorate when the ductus closes, so early detection prevents avoidable deaths. Newborn pulse-ox screening (right hand and one foot at 24-48 h) identifies hypoxemia that exam can miss. Act fast: refer if SpO_2 <90% anytime, or if repeat readings remain 90-94% or differ by >3%; confirm with echocardiography.

- Acyanotic (left-to-right shunts): atrial septal defect (ASD), ventricular septal defect (VSD), patent ductus arteriosus (PDA), atrioventricular septal defect (AVSD).
- Cyanotic (right-to-left shunts): Tetralogy of Fallot (TOF), transposition of the great arteries (TGA), tricuspid atresia, total anomalous pulmonary venous return (TAPVR).
- Obstructive: pulmonary stenosis (PS), aortic stenosis (AS), coarctation of the aorta (CoA).

Screening for critical congenital heart disease at birth

Newborn Screening Protocol for Critical Congenital Heart Disease (CCHD)

Target population

Screen all newborns ≥24 hours old in well-baby areas prior to discharge. Screen earlier only if discharge is planned before 24 h. Symptomatic infants or those with known/suspected CHD go directly to clinical/cardiology evaluation. (American Academy of Pediatrics)

Timing

- Preferred: at ≥24 hours of life (reduces false positives).
- If discharge <24 h: screen as late as feasible before discharge. Infant should be on room air. (American Academy of Pediatrics)

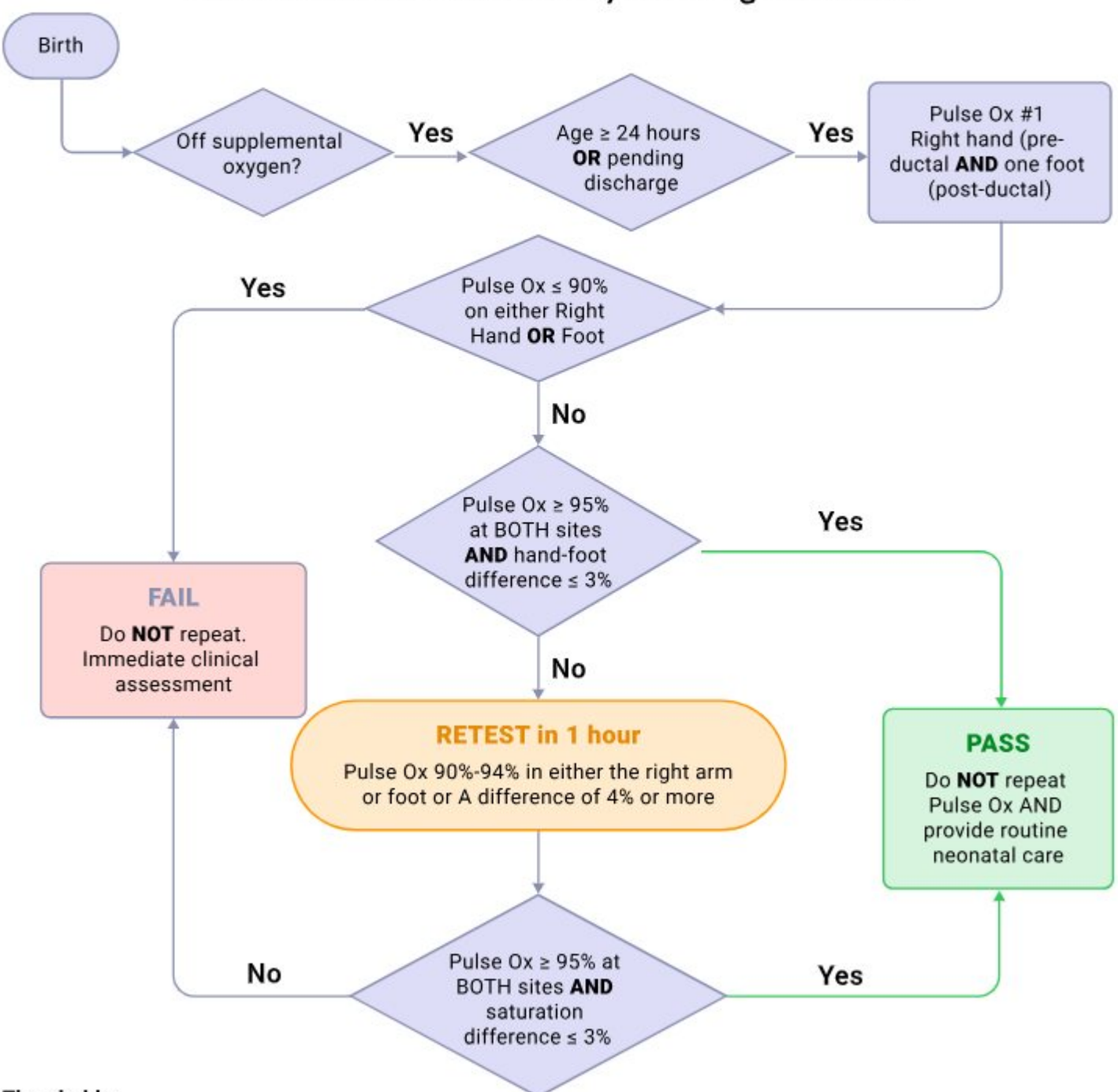
Equipment & site

- Motion-tolerant neonatal pulse oximeter with appropriate probe.
- Measure pre-ductal saturation on right hand and post-ductal on either foot (simultaneously if two devices; sequentially if one). (American Academy of Pediatrics)

Procedure

- Ensure infant is warm, calm, and well-perfused; verify probe placement and signal quality.
- Record SpO_2 (RH and foot) to nearest whole percent.
- Document vitals and any clinical concerns. (American Academy of Pediatrics)

Newborn CCHD Pulse-Oximetry screening - Flowchart



Thresholds:

PASS = both ≥ 95% with ≤ 3% difference

IMMEDIATE FAIL = any value ≤ 90% (either site any time) -Urgent evaluation;

BORDERLINE = 90-94% or ≥ 3% difference - repeat once in 1 hour.

CCHD essentials for screen-positive infants

- Target lesions (early clues - take immediate step)
- TGA: cyanosis not improving with O_2 - start PGE_1, urgent echo.
- HLHS / critical coarctation/IAA: shock days 2-7, weak femoral - give PGE_1, 4-limb BP, echo, urgent transfer.
- Pulmonary atresia/critical PS: profound cyanosis – give PGE_1, echo.
- TAPVR (obstructed): cyanosis + distress - stabilize, urgent surgery after echo.
- Critical ToF: cyanosis/spells - consider PGE_1 if severe RVOTO, echo.
- Truncus arteriosus / tricuspid atresia: cyanosis ± heart failure - echo, supportive care.

"Do not miss" signs (act even if screen passed)

SpO_2 <90%, persistent 90-94% or >3% difference after repeat, central cyanosis unresponsive to O_2, shock/acidosis day 2-7, weak femoral pulses, unexplained tachypnea/poor feeding/lethargy.

Interpretation (AAP/CDC algorithm)

PASS: Both RH and foot ≥95% and difference ≤3%.

IMMEDIATE FAIL: Any reading <90% (either site) at any time → urgent evaluation.

BORDERLINE: Any 90-94% or RH-foot difference >3% → repeat in 1 hour; up to 2 total screens. If still borderline after the second screen → FAIL. (American Academy of Pediatrics)

Actions after a FAILED screen

Urgent clinical assessment (airway, breathing, circulation; check for sepsis/PPHN/pneumonia). (details see below)

Echocardiography where available; initiate oxygen/supportive care per neonatal protocol; arrange referral to designated tertiary center (enter local pathway/contacts). (CDC)

Documentation

Age at test, RH and foot SpO_2, difference, pass/fail, repeats, actions taken, clinician signature; file in newborn record and electronic system (if available). (CDC)

Onset of Cyanosis by Age

Onset window	Usual bedside picture	Likely lesions (examples)
Day 1-7 (first week)	Deep cyanosis, relatively well-perfused; minimal distress	d-TGA with intact ventricular septum
Day 1-7 (first week)	Shock/poor pulses; metabolic acidosis	Duct-dependent systemic flow: HLHS, critical AS, interrupted aortic arch/coarctation
Day 1-7 (first week)	Severe cyanosis without much distress; clear lungs	Pulmonary atresia, critical PS, severe TOF physiology, Ebstein anomaly
Day 1-7 (first week)	Cyanosis with respiratory distress; loud P2/ wet lungs	Obstructed TAPVC
Day 1-7 (first week)	Cyanosis with high pulmonary blood flow signs	Truncus arteriosus
7 days-1 month	Persistent cyanosis beyond immediate neonatal period	d-TGA (± VSD), severe PS, TOF, truncus arteriosus
Late infancy/ childhood	Cyanosis appears after prior VSD physiology	TOF, DORV with PS, d-TGA with VSD+PS, tricuspid atresia with VSD+PS

Abbrev: d-TGA dextro-transposition of the great arteries; HLHS hypoplastic left heart syndrome; AS aortic stenosis; PS pulmonary stenosis; TOF tetralogy of Fallot; DORV double-outlet right ventricle; TAPVC total anomalous pulmonary venous connection; VSD ventricular septal defect.

Neonate with Possible Congenital Heart Disease (CHD):

Ask / Look / Feel	Hemodynamic interpretation	Examples of CHD to consider	First steps
Decreased activity; poor feeding	Low cardiac output or evolving heart failure/ shock	Duct-dependent systemic flow: HLHS, critical aortic stenosis (AS), interrupted aortic arch/ coarctation; also large shunt lesions once PVR falls	Assess perfusion, glucose, ABG; start prostaglandin E1 (PGE1) if duct-dependent lesion suspected; urgent echo/ cardiology consult
Cyanosis on exam or low SpO_2 on pulse oximetry (pre/ post-ductal)	Central cyanosis from: (1) reduced pulmonary blood flow; (2) mixing lesion with high pulmonary flow; or (3) pulmonary venous obstruction	Reduced PBF: pulmonary atresia, critical PS, severe TOF, Ebstein; Mixing/high PBF: d-TGA (± VSD), truncus; Venous obstruction: obstructed TAPVC	Confirm pre/post-ductal saturations; give PGE1 if duct-dependent physiology possible; avoid excessive oxygen in mixing lesions; urgent echo
Respiratory distress / tachypnea	Pulmonary overcirculation (L→R shunt or mixing with high PBF) or pulmonary venous hypertension/ obstruction	Large VSD/PDA/AVSD; d-TGA with VSD; truncus; obstructed TAPVC (if severe distress with loud P2 and edema)	CXR, ABG; diuretics/respiratory support as indicated; urgent echo to differentiate overcirculation vs venous obstruction
Inappropriate tachycardia or bradycardia for age	Arrhythmia with hemodynamic compromise → low output/shock	SVT, complete heart block (may coexist with CHD)	ECG; treat arrhythmia per protocol (vagal/adenosine for SVT, pacing for CHB); support perfusion

Signs of shock: cold clammy peripheries, CRT >3 s, core-peripheral gap >2°C	Duct-dependent systemic circulation or severe ventricular dysfunction	HLHS, critical AS, interrupted aortic arch/coarctation; severe myocarditis/ventricular dysfunction (differential)	Airway/breathing; cautious fluids; start PGE1; correct acidosis; inotropes as needed; urgent echo and transfer
Tachypnea, tachycardia, hepatomegaly (heart failure signs)	Left-to-right shunt with pulmonary overcirculation once PVR falls	Large VSD, PDA, AVSD	Diuretics/afterload management; feeding support; echo and cardiology review
Feeding pattern abnormal (fatigue, diaphoresis, poor weight gain)	May reflect heart failure, low output, or significant desaturation with feeds	As above- large shunts; duct-dependent systemic lesions; mixing lesions with low systemic saturation	Weigh, monitor feeds and saturations; evaluate for HF/desaturation; escalate per findings and obtain echo

Abbrev: CHD congenital heart disease; HLHS hypoplastic left heart syndrome; AS aortic stenosis; PVR pulmonary vascular resistance; PBF pulmonary blood flow; PS pulmonary stenosis; TOF tetralogy of Fallot; d-TGA dextro-transposition of the great arteries; VSD ventricular septal defect; TAPVC total anomalous pulmonary venous connection; PDA patent ductus arteriosus; AVSD atrioventricular septal defect; SVT supraventricular tachycardia; CHB complete heart block; PGE1 prostaglandin E1; ABG arterial blood gas; CXR chest X-ray; CRT capillary refill time.

Drug -Dosages

Indication	Drug (name)	Dose	Route	Duration	Cautions
Duct-dependent systemic/ pulmonary flow	Prostaglandin E_1 (alprostadil)	0.05-0.1 µg/kg/min → titrate to 0.01-0.025	IV infusion	Until definitive therapy	Apnea, hypotension; airway ready
PDA closure (preterm)	Paracetamol	15 mg/kg q6h	PO/IV	3-7 days	LFTs; hepatic disease
	Ibuprofen	10 mg/kg, then 5 mg/kg at 24 & 48 h	PO/IV	3 doses	Renal/GI caution

Initial stabilization bundle (suspected CCHD)

SHOCK
Poor feeding, lethargy Tachypnea, Cyanosis, Cold extremities, Weak pulses,
Prolonged capillary refill, hypotension, Metabolic acidosis

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Airway—breathing—circulation; keep warm; NPO.
IV/IO access and fluid resuscitation: 10 ml/kg isotonic fluid (max 40 ml/kg). Stop if hepatomegaly develops.
Correct hypothermia, hypoglycemia, hypocalcemia.
Correct acidosis, Start antibiotics.
Monitor response. Avoid NSAIDs/indomethacin.

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Manage according to neonatal shock guidelines

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Keep high index of suspicion & refer early—even with minimal signs or uncertainty—for specialist assessment.

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Refractory shock, unlikely sepsis → Urgent referral to pediatric cardiologist

Start PG E1 (0.05—0.1 µg/kg/min (titrate 0.01—0.1) & refer if:

- Femoral pulses markedly weaker than upper limb pulses
- Right arm— foot SpO_2 difference
- Arrange echo and urgent transfer; transport on PGEI with monitoring.

Septic shock likely if:

- Predisposing maternal and neonatal factors
- Core peripheral temperature difference > 3
- Sepsis Screen Positive (CRP/TLC/ANC/Platelets abnormal; blood culture sent)

Neonatal Cyanosis (Bed-side algorithm)

Start: Cyanosis with peripheral oxygen saturation (SpO_2) <95% (room air)

1. Stabilize + basics

- Airway-Breathing-Circulation, warm, glucose, IV access, continuous SpO_2.
- Obtain pre/post-ductal SpO_2 if possible.

2. Chest X-ray (CXR) to screen for obstructed pulmonary venous return

- **Suggestive CXR**: suspect obstructed total anomalous pulmonary venous connection (TAPVC) → immediate referral to paediatric cardiology/cardiothoracic center.
- If not suggestive → proceed.

3. Assess severity by saturation

- **SpO_2 <80% (or falling)**: think duct-dependent pulmonary circulation (e.g., pulmonary atresia/critical pulmonary stenosis [PS]) or transposition of the great arteries (TGA) with intact septum.
  - Action now: Start prostaglandin E_1 (PGE_1; alprostadil) infusion and arrange urgent paediatric cardiology evaluation within hours.
  - be prepared for apnoea; airway support may be needed.
- SpO_2 ≥80%:
  - Early paediatric cardiology consult + echocardiography.
  - Delay discharge until diagnosis confirmed.
  - Close SpO_2 monitoring during the first week (watch for falls with ductal closure).
  - **If only heart-failure features present: start heart failure (HF) management as indicated; consider iron supplementation.**

4. Ongoing trigger

- If SpO_2 <80% on serial monitoring at any point → start PGE_1 and escalate urgently as above.

Decision point	Findings	Next action
CXR suggests obstructed pulmonary venous return and RDS unlikely	"White-out"/ground-glass, pulmonary venous hypertension	Suspect obstructed TAPVC → immediate referral
Saturation very low or dropping	SpO_2 <80% (initial or serial)	Suspect duct-dependent lesion or TGA (intact septum) → Start PGE_1 (0.05-0.1 µg/kg/min IV), titrate 0.01-0.1 µg/kg/min to effect; urgent paediatric cardiology

Saturation not in critical range	SpO_2 ≥80%	Early cardiology + echo, close first-week SpO_2 monitoring; manage HF if present; consider iron
Any path along the way	Significant CHD suspected	Keep NPO if unstable, secure transport to cardiac center, maintain thermoneutrality and glucose

For pediatric heart failure see section II on congenital heart disease in Children

SECTION II

CONGENITAL HEART DISEASE IN CHILDREN: APPROACH TO MANAGEMENT

- Clinical features: cyanosis, heart failure (tachypnea, poor feeding/failure to thrive (FTT), syncope/exercise intolerance, recurrent pneumonia, cyanotic spells.
- Exam: pulses (radio-femoral delay), BP all limbs if CoA suspected, pre/post-ductal SpO_2, hepatomegaly, edema; murmurs (e.g., VSD pansystolic left lower sternal border (LLSB); ASD ejection left upper sternal border (LUSB) with fixed split S2; PDA continuous left infraclavicular).
- Initial tests: ECG, CXR, pulse-ox (consider critical CHD (CCHD) screen in newborns).
- Confirm: echocardiography (2D/ Doppler; 3D/MRI/CT if complex); cardiac catheterization for hemodynamics when needed.

Left to Right Shunts are the most common type of congenital heart defects and is one of the common causes of infant morbidity and mortality. Majority of the lesions are easily correctable if detected on time.

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

Suspected Left-to-Right Shunt (Pediatric)

Table 1. When to suspect?

Criterion	Details
Failure to thrive	Weight <3rd centile for age or drop by ≥2 major centile lines
Feeding difficulty with cold sweats	Suck-rest-suck cycle; forehead sweating
Recurrent infections	Repeated chest infections or one life-threatening episode
Baseline tachypnea with retractions	RR >60/min (<1 year) or >50/min (1-2 years); subcostal/intercostal indrawing
Tachycardia	HR >160/min (<1 year) or >140/min (1-2 years)
Bounding pulses	High-volume pulse (typical of PDA/AP window)
Precordial changes	Precordial bulge with active precordium
Auscultation	Loud S2, gallop; ejection systolic or mid-diastolic murmurs (note: very large shunts may have soft murmurs)
Hepatomegaly	Congestive hepatomegaly on palpation
Dysmorphism	Down syndrome → consider AVSD
Peripheral pulses	Feeble lower-limb pulses → consider CoA

Table 2. Investigations (Essential first-line)

Test	Purpose	Notes
Chest X-ray (CXR)	Cardiac silhouette, pulmonary vasculature, PA dilation, skeletal anomalies	
Electrocardiogram (ECG)	Rate/rhythm, chamber enlargement, unexpected abnormalities	Screen for tachyarrhythmia; RS complexes; ischemia patterns when relevant
Blood tests	CBC and electrolytes based on clinical context	Add BNP/NT-proBNP if available per protocol
Echocardiography	Confirms shunt lesion, size, sites; assesses chamber dilation, pulmonary pressures, AV valve regurgitation	Large post-tricuspid shunts require **early referral**

Modified Ross classification for Heart failure in children

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

Age-specific normal vitals

Age group	Heart rate (awake, bpm)	Respiratory rate (/min)	Systolic BP (mmHg)	SpO_2 (room air)
Neonate (0-28 d)	100-205	40-60	~60-76	≥95%
Infant (1-12 mo)	100-180	30-53	72-104	≥95%
Toddler (1-3 y)	98-140	22-37	86-106	≥95%
Preschool (4-5 y)	80-120	20-28	89-112	≥95%
School-age (6-12 y)	75-118	18-25	97-115	≥95%
Adolescent (13-18 y)	60-100	12-20	110-131	≥95%

Use the vitals table to interpret symptom severity (tachycardia, tachypnea, hypotension) for the child's age when applying the severity criteria above.

Table 3. Referral

When to refer
Timely referral to a higher center with pediatric cardiac services; large post-tricuspid shunts (e.g., large VSD/PDA/AVSD) warrant **early** referral.

Differential diagnosis

Respiratory disease (RDS, pneumonia, bronchiolitis, persistent pulmonary hypertension of the newborn (PPHN), sepsis, metabolic acidosis, innocent/ functional murmurs, cardiomyopathy, myocarditis, rheumatic/degenerative valve disease.

Management goals & principles

- Stabilize circulation; relieve symptoms; prevent irreversible pulmonary vascular disease; correct repairable anatomy; prevent arrhythmias/ endocarditis; ensure growth and good quality of life; plan transition to adult congenital heart disease (ACHD).

Table 4. Approach to CHD Management by Type & Age

CHD Type	Examples	Initial/Supportive Management	Definitive / Long-Term Management
Acyanotic CHD - Infants	ASD, VSD, PDA, AVSD	▪ Treat heart failure if symptomatic (diuretics, optimize feeding) ▪ Monitor growth and development ▪ PDA in preterm: paracetamol/ ibuprofen/indomethacin per protocol	▪ ASD: close if RA/RV dilation or Qp:Qs ≥1.5 (device if secundum, surgery otherwise) ▪ VSD: observe if small; close if moderate/ large with symptoms, LV dilation, or aortic cusp prolapse ▪ PDA: device closure in term infants ▪ AVSD: early complete repair at 3-6 months if complete form; address AV valve regurgitation

Acyanotic CHD - Children/ Adolescents	Small VSD/ASD, partial AV canal defects	▪ Monitor for spontaneous closure ▪ Address heart failure ▪ Assess growth, exercise tolerance	▪ Elective closure if hemodynamically significant or symptomatic
Cyanotic CHD - Neonates/ Infants	TOF, TGA, TAPVC, Tricuspid atresia, Pulmonary atresia, Critical PS, HLHS	▪ Stabilize airway and breathing ▪ PGE_1 infusion to maintain ductal patency (Dose see above) ▪ Balloon atrial septostomy if TGA with restrictive atrial septum ▪ Urgent referral to tertiary cardiac center	▪ TOF: manage spells acutely; complete repair at 3-6 months (earlier if recurrent spells) ▪ TGA: arterial switch in neonatal period ▪ TAPVC: urgent surgery if obstructed; early repair otherwise ▪ CoA: PGE_1 in neonates; surgical repair in neonates or balloon/stent in older children; long-term HTN control
Cyanotic CHD - Children/ Adolescents	Repaired or unrepaired TOF, Eisenmenger physiology	▪ Assess oxygen saturation and cardiac function ▪ Manage complications (polycythemia, hemoptysis, arrhythmias, cyanotic spells in TOF physiology, heart failure in lesions with increased pulmonary flow)	▪ Surgical/catheter interventions where feasible ▪ Eisenmenger: supportive/pulmonary vasodilator therapy, avoid pregnancy
Single-Ventricle Physiology	HLHS, tricuspid atresia with inadequate RV, unbalanced AV canal	▪ Balance systemic vs pulmonary blood flow (Qp:Qs) ▪ Interstage monitoring after stage I surgery	▪ Staged palliation: Norwood → Glenn → Fontan ▪ Lifelong surveillance for arrhythmia, protein-losing enteropathy, ventricular dysfunction
CHD in Adults (ACHD)	Repaired complex CHD, residual shunts, pulmonary hypertension	▪ Regular follow-up at ACHD/ cardiology clinic ▪ Manage arrhythmias, heart failure, pulmonary vascular disease	▪ Lifelong specialist follow-up ▪ Surgical or catheter re-interventions as required

Note:
- Advice patient to avoid pregnancy if Eisenmenger syndrome develops as it carries very high maternal and fetal mortality.
- Pulse-ox screening; standardized echo views; tele-cardiology for remote review.
- Transport protocols; early referral triggers; nutrition support; family education.

Table 5. GENERAL ADVICE TO CAREGIVERS

Topic	Key messages
Infection prevention	Educate about hygiene to prevent infections
Feeding	Promote breastfeeding if tolerated; teach gavage/spoon feeds if breastfeeding is difficult - prefer expressed breast milk
Top-up feeds	Use top milk/formula if breast-milk output low; target ~120 mL/kg/day total intake
Weaning foods (>6 months)	Use energy-dense complementary feeds
Immunization	Continue as per IAP schedule
Supplements	Vitamin D3, calcium, iron per IAP recommendations and clinical need

Pharmacological therapy

Indication	Drug (name)	Dose	Duration	Cautions
Cyanotic spell	Propranolol (PO)	1-4 mg/kg/day in 3 divided doses	Long term	Hypotension
Heart failure (peds)	Furosemide (Po/IV)	1-2 mg/kg q12-24h (max 2 mg/kg/dose); (neonate 0.5-1 mg/kg); refractory infusion 0.05-0.2 mg/kg/h q 8-12 h oral suspension 10 mg/mL → 0.1 mL/kg/dose	As needed; reduce/hold during diarrhea or vomiting;	Monitor UO, weight, electrolytes (Na^+/K^+/Mg^{2+}), creatinine; ototoxicity with rapid IV
	Add if loop diuretic > once daily			
	Spironolactone (PO)	1-3 mg/kg/day ÷ q12-24h	Long-term adjunct	K^+/Cr
	Enalapril (PO)	0.05-0.1 mg/kg/dose q12-24h (adult: 2.5-5 mg bid start)	Long-term	BP, K^+, Cr; avoid early neonates/AKI
	Digoxin (PO/IV)	5-10 µg/kg/day ÷ q12h (optional load 30-50 µg/kg total)	Long-term	Levels if concern; arrhythmia, renal dosing
Pulmonary hypertension	Sildenafil (PO)	0.5-2 mg/kg q6-8h (adult 20 mg tid)	Long term	Hypotension; avoid nitrates
	Bosentan (PO)	2 mg/kg q12h (adult 62.5→125 mg bid)	Long-term	Monthly LFTs; teratogenic
Arrhythmias	Amiodarone (IV/PO)	Load 5-10 mg/kg IV, then 5-15 mg/kg/day PO	Individualize	Thyroid/LFTs/QT
	Metoprolol (PO)	0.5-1 mg/kg q6-12h (max 200 mg/day)	Long-term	Bradycardia, hypotension
Antiplatelet/ anticoagulants	Aspirin (PO)	3-5 mg/kg/day (adult 75-100 mg/day)	Long-term	GI bleed risk
	Warfarin (PO)	Per INR 2-3	As indicated	INR checks, interactions
Infective endocarditis (IE) prophylaxis (high-risk)	Amoxicillin (PO)	50 mg/kg (max 2 g) 30-60 min pre-dental	Single dose	Allergy: cephalexin; if allergic azithromycin/ clarithromycin. *Clindamycin not recommended*

For details of management of heart failure and arrhythmia see respective guidelines.

Assessment of response & follow-up

Item	Criteria / Actions
Improving	▪ Better feeding/exercise; stable weight/growth ▪ HR/BP/SpO_2 at target; BNP falling ▪ Echo: improved function, less regurgitation/gradient
Worsening	▪ Tachycardia/bradycardia for age; hypotension or hypertension ▪ Drop in SpO_2; edema/hepatomegaly; BNP rising ▪ New arrhythmia; lower EF on echo
Adjustment interval	▪ Adjust therapy every 2-4 weeks during titration; then every 3-6 months ▪ Step-up for symptoms/echo worsening; step-down cautiously after sustained stability
Referral - Emergency	▪ Refractory cyanosis/hypoxemia, shock/severe HF, refractory arrhythmia, critical lesions at birth ▪ Start PGE_1 if indicated and transfer to tertiary center
Referral Urgent (24-72 h)	▪ New moderate-severe CHD with symptoms; rising PAP; progressive valve disease ▪ Rapid pediatric cardiology review
Referral - Routine	▪ Stable repaired lesions or mild CHD ▪ Schedule ACHD/pediatric cardiology follow-up
Documentation (before transfer)	▪ Record all pre-referral interventions: drug name-dose-route-time; oxygen modality/ FiO_2; lines; key tests; clinical response

Complications

Arrhythmias, heart failure, pulmonary hypertension, endocarditis, thromboembolism, residual/recurrent lesions, Eisenmenger, growth failure, pregnancy risk in ACHD, Fontan-associated issues.

Patient education: objectives & instructions

- Know the diagnosis/plan, meds and dosing, adherence, side-effects.
- Dental hygiene and IE prophylaxis rules.
- Vaccines: influenza, pneumococcal, COVID-19; RSV where applicable.
- Activity guidance: what's allowed/ restricted; hydration; avoid high altitude if advised.
- Red flags: increased cyanosis/SOB, syncope, chest pain, rapid weight gain/ edema, fever.
- Follow-up: keep appointments; bring weight/SpO_2 logs.
- Family planning: contraception/ pregnancy counselling in ACHD.

SECTION III

INTRODUCTION

Congenital heart disease (CHD) is a spectrum of structural abnormalities of the heart and great vessels present from birth, caused by disordered fetal development; lesions range from trivial to life-threatening. It excludes inherited cardiomyopathies or syndromes (such as Marfan syndrome or hypertrophic cardiomyopathy) and normal variants like a patent foramen ovale. Congenital valvular disease (for example, a bicuspid aortic valve) often follows general valve-disease guidance, with congenital nuances.

Globally, CHD affects ~8-12 per 1,000 live births, and modern care now allows more than 90% of affected children to reach adulthood, creating a growing adult congenital heart disease (ACHD) population that still faces lifelong risks of arrhythmias, heart failure, pulmonary hypertension, residual or recurrent lesions, and re-intervention. In Southeast Asia, incidence is similar, but outcomes are limited by delayed detection and constrained surgical access; where in-country surgery is scarce, overseas referral is common, making neonatal screening and reliable pediatric follow-up crucial. Many lesions present in infancy and some require urgent repair (e.g., large ventricular septal defects or complex cyanotic CHD), while adults may present with repaired or unrepaired defects and late sequelae.

Lifelong follow-up remains essential even after "complete" repair. Common pitfalls include late referral, weak follow-up, poorly timed procedures, managing non-cardiac surgery without ACHD expertise, and empirical drug use without confirming anatomy or hemodynamics. Standardized pathways, early detection, correct triage, pre-referral stabilization, and consistent follow-up reduces preventable morbidity and death.

SCOPE OF THE GUIDELINES

Practical, evidence-based care for common CHD in children and adults - screening, diagnosis, stabilization, and follow-up. Heart failure, arrhythmia, surgical and Electrophysiology details are referenced but details not included. Lesion-specific, age group specific strategies highlight differences from acquired disease. Usable from primary to secondary care, until tertiary intervention is available.

Users. Primary care doctors, pediatricians, internists, emergency physicians, cardiologists, nurses, paramedics, community health workers and policy/program leads.

Roles by level.

- **Primary**: early detection (antenatal, neonatal, school health), high clinical suspicion, ECG/CXR ± basic echo, start stabilization, arrange urgent referral. CCHD screening as per Maldives' National protocols provided.

- **Secondary**: confirm diagnosis with echo, optimize medical therapy, prepare for surgery/catheter intervention, manage uncomplicated cases.
- **Tertiary**: complex surgery, advanced catheter work, complication management, specialized heart-failure care.

Bridging & pathways. Emphasize early stabilization, safe transport, clear urgent vs elective pathways, simplified protocols for limited diagnostics, and standard handover templates to ensure that care is timely, safe, and coordinated across the system.

DEFINITIONS

CHD encompasses a wide range of structural cardiac abnormalities present from birth, resulting from disruptions in normal fetal heart development. Understanding these categories such as acyanotic and cyanotic lesions, obstructive defects, and complex anomalies is essential for accurate diagnosis, appropriate triage, and targeted treatment planning.

Category	Type	Definition
Acyanotic CHD - Left-to-Right Shunt	Atrial Septal Defect (ASD)	Defect in the interatrial septum allowing blood flow from left atrium to right atrium.
	Ventricular Septal Defect (VSD)	Opening in the interventricular septum causing blood to flow from left ventricle to right ventricle.
	Patent Ductus Arteriosus (PDA)	Persistent fetal connection between the aorta and pulmonary artery.
	Atrioventricular Septal Defect (AVSD)	Failure of endocardial cushion fusion, causing combined ASD and VSD with common AV valve.
Acyanotic CHD - Obstructive Lesions	Pulmonary Stenosis (PS)	Narrowing at or near pulmonary valve causing RV outflow obstruction.
	Aortic Stenosis (AS)	Narrowing of the aortic valve or LV outflow tract.
	Coarctation of the Aorta	Constriction of the aorta, distal to left subclavian artery.
Cyanotic CHD - Decreased Pulmonary Blood Flow	Tetralogy of Fallot (TOF)	Combination of VSD, pulmonary stenosis, RV hypertrophy, and overriding aorta.
	Tricuspid Atresia	Absence of tricuspid valve with hypoplastic RV and obligatory interatrial communication.
Cyanotic CHD - Increased Pulmonary Blood Flow / Mixing Lesions	Transposition of the Great Arteries (TGA)	Aorta from RV, pulmonary artery from LV, causing parallel circulation.
	Total Anomalous Pulmonary Venous Connection (TAPVC)	Pulmonary veins drain into right atrium or systemic veins instead of left atrium.
Complex CHD	Hypoplastic Left Heart Syndrome (HLHS)	Underdevelopment of left heart structures including LV, mitral valve, aortic valve.
	Double Outlet Right Ventricle (DORV)	Both great arteries arise from RV, often with associated VSD.

CAUSES, RISK FACTORS & TRIGGERS

Domain	Subcategory	Examples
Causes	Abnormal fetal cardiac morphogenesis	Malformation during the first 8 weeks of gestation
	Multifactorial	Genetic, environmental, teratogenic influences
Risk factors	Genetic	Chromosomal: Trisomy 21, 18, 13; single-gene defects
	Maternal	Diabetes, phenylketonuria, rubella, teratogens: retinoic acid, alcohol, lithium
	Family history	Higher recurrence risk in siblings/first-degree relatives
Triggers for symptomatic presentation	Intercurrent illness	Infection (e.g., pneumonia), fever, dehydration
	Hematologic/arrhythmic	Anemia, arrhythmias
	Physiologic stress	High altitude/hypoxia
	Reproductive	Pregnancy in undiagnosed or partially repaired CHD

EVALUATION FOR DIAGNOSIS

Evaluation of Suspected Congenital Heart Disease

Domain	Key items	What to look for
History	Antenatal	Maternal illnesses (diabetes, infections), teratogen exposure; anomaly scan 18-22 weeks and results
	Birth	Cyanosis at birth, low Apgar, resuscitation, prolonged NICU stay
	Feeding/growth	Poor feeding, diaphoresis with feeds, failure to thrive
	Exercise tolerance	Tachypnea, dyspnea on exertion, syncope, chest pain in older child
	Respiratory infections	Recurrent LRTIs, wheeze, prolonged cough
Physical exam	General	Cyanosis, clubbing, growth parameters and centiles
	Cardiovascular	BP in all limbs if indicated, pulses (radio-femoral delay), murmurs, added sounds, heave; signs of heart failure (use age-specific vitals and HF signs list)
	Respiratory	Work of breathing, crackles if pulmonary edema/ congestion

Domain	Key items	What to look for
Initial investigations	ECG	Rhythm/arrhythmia, chamber hypertrophy, axis deviation
	Chest X-ray	Cardiomegaly, pulmonary vascular markings, cardiac silhouette
	Pulse oximetry	Baseline SpO_2; perform CCHD screen if not already done
Targeted investigations	Echocardiography	Primary tool for diagnosis and follow-up; 2D echo defines anatomy and hemodynamics. 3D echo (if available) adds detailed morphology and valve assessment, helpful in complex lesions and surgical planning
	Cardiac MRI / CT	Define complex anatomy, extracardiac vessels, ventricular volumes and function; useful post-surgery or when echo is limited
	Cardiac catheterization	Hemodynamics (pressures, shunts, PVR), coronary anatomy; pre-intervention planning or when noninvasive data are inconclusive
	Genetic testing	If syndromic features, extracardiac anomalies, or family history suggest a genetic etiology

DIFFERENTIATE FUNCTIONAL MURMURS FROM PATHOLOGICAL MURMURS

Functional murmur i.e., non-pathological heart murmurs produced by normal blood flow through structurally normal valves and chambers. They are common in children, adolescents, pregnancy, fever, anemia, or hyperthyroidism where cardiac output is increased. Distinguishing functional from pathological murmurs prevents unnecessary tests and anxiety while ensuring early detection of structural heart disease. Functional murmurs need only reassurance, whereas pathological murmurs require investigation and referral and efficient care is critical for better outcomes.

Table contrasting functional vs pathological murmurs

Feature	Functional (Innocent/Physiologic) Murmurs	Pathological Murmurs
Timing	Short, systolic ejection (early-mid systolic)	Can be systolic, diastolic, continuous; holosystolic or diastolic especially suspicious
Intensity	Soft (≤ grade 2/6), often variable	Loud (≥ grade 3/6), may be harsh, with thrills
Quality	Blowing, musical, vibratory	Harsh, rumbling, or associated with clicks/ snaps

Location	Left sternal border, pulmonary area	May be localized to apex, aortic, or other areas; often radiates (to neck, axilla, back)
Radiation	None	Often present (e.g., mitral regurg → axilla, aortic stenosis → carotids)
Variation with posture	Changes with position (softer upright, louder supine), may vary with respiration	Usually consistent; does not disappear with position change
Associated findings	No symptoms; normal pulses, BP, growth; normal S2	May have abnormal S2, clicks, heaves, cyanosis, heart failure signs, poor growth/ exercise intolerance
Clinical context	Common in children, adolescents, pregnancy, fever, anemia, hyperthyroidism	Associated with structural heart disease, valvular lesions, congenital defects
Need for investigation	Usually none if classical features; reassurance only	Requires further evaluation (ECG, echocardiography, specialist referral)

CONFIRMATION OF DIAGNOSIS

Accurate confirmation relies on integrating history, physical examination, and targeted investigations, with echocardiography as the primary diagnostic tool. Additional imaging, cardiac catheterization, and functional tests may be required for complex lesions. A precise diagnosis not only guides immediate management but also determines the timing and type of intervention, long-term follow-up needs, and prognosis.

CHD Type	Key Diagnostic Features on Echo	Supportive Findings
ASD	Defect in atrial septum; left-to-right shunt	Systolic ejection murmur at LUSB from ↑ pulmonary flow; fixed split S2; RA/RV enlargement
VSD	Defect in ventricular septum with shunt	Harsh pansystolic murmur at LLSB ± thrill; LV volume overload; possible hepatomegaly/failure to thrive if large
PDA	Continuous flow between aorta and PA	Continuous "machinery" murmur at left infraclavicular area; bounding pulses, wide pulse pressure; LA/LV enlargement if significant
TOF	VSD + RV outflow obstruction + overriding aorta + RVH	Harsh systolic ejection murmur at LUSB (RVOT); cyanosis/spells; boot-shaped heart on CXR
TGA	Parallel great vessel origin	Often no murmur; single loud S2; severe cyanosis in newborn; murmur only if VSD/PS present
Coarctation	Narrowed aortic segment	Ejection systolic murmur over left infrascapular/back; BP arm-leg difference >20 mmHg, radio-femoral delay; possible LVH

CLASSIFICATION & SEVERITY ASSESSMENT

A structured framework to categorize congenital heart disease based on anatomical type, physiological impact, and clinical status helps prioritize urgency, determine appropriate management strategies, and guide referral decisions.

Classification by Pathophysiology

Pathophysiology class	Representative lesions	Core physiology & presentation	Auscultation highlights
Acyanotic CHD (left-to-right shunts)	Ventricular septal defect (VSD), Atrial septal defect (ASD), Patent ductus arteriosus (PDA), Atrioventricular septal defect (AVSD)	Oxygenated blood shunts left→right, increasing pulmonary blood flow. Common features: murmur, recurrent chest infections, heart failure if large shunt, failure to thrive.	▪ VSD: harsh pansystolic murmur at LLSB ± thrill. ▪ ASD (secundum): ejection systolic murmur at LUSB with fixed split S2 (↑ pulmonary flow). ▪ PDA: continuous "machinery" murmur at left infraclavicular area; bounding pulses, wide pulse pressure. ▪ AVSD: holosystolic murmur from AV valve regurgitation at LLSB/apex; possible flow murmur at LUSB.
Cyanotic CHD (right-to-left shunts)	Tetralogy of Fallot (TOF), Transposition of the Great Arteries (TGA), Tricuspid atresia, Total anomalous pulmonary venous return (TAPVR)	Deoxygenated blood enters systemic circulation → central cyanosis, clubbing, exercise intolerance.	▪ TOF: harsh ejection systolic murmur at LUSB from RVOT obstruction. ▪ TGA: often no murmur; murmur only if VSD or pulmonary stenosis present; single loud S2 common. ▪ Tricuspid atresia: usually no ejection murmur; VSD holosystolic murmur may be present; single S2. ▪ TAPVR: murmur often absent; obstructed forms present with respiratory distress rather than a loud murmur.
Obstructive lesions	Pulmonary stenosis (PS), Aortic stenosis (AS), Coarctation of the aorta (CoA)	Outflow obstruction raises ventricular workload; may cause heart failure. CoA often presents with upper-limb hypertension and arm-leg BP gradient.	▪ PS: ejection systolic murmur at LUSB with ejection click; may radiate to the back. ▪ AS: ejection systolic murmur at RUSB radiating to carotids; soft/absent A2 if severe. ▪ CoA: ejection systolic murmur best heard interscapular/back; radio-femoral delay and arm-leg BP gradient.

CHD congenital heart disease; LLSB left lower sternal border; LUSB left upper sternal border; RUSB right upper sternal border; S2 second heart sound; AV atrioventricular; RVOT right-ventricular outflow tract; BP blood pressure; VSD ventricular septal defect; ASD atrial septal defect; PDA patent ductus arteriosus; AVSD atrioventricular septal defect; TOF tetralogy of Fallot; TGA transposition of the great arteries; TAPVR total anomalous pulmonary venous return; PS pulmonary stenosis; AS aortic stenosis; CoA coarctation of the aorta.

Severity Assessment

Severity in congenital heart disease (use with age-specific vitals below)

Domain	Thresholds / Examples	How to assess
Hemodynamic impact	Large shunt Qp:Qs >2:1; severe gradients >50 mmHg; pulmonary hypertension	Echo/Doppler, cardiac catheterization if needed
Symptom burden	NYHA II-IV (or pediatric equivalent); failure to thrive	History, growth chart, functional class

Oxygen saturation	Severe cyanosis SpO_2 <85% (room air)	Pre/post-ductal pulse oximetry
Functional status	Reduced 6-min walk distance; impaired CPET	6MWT/CPET vs norms
Complications	Arrhythmias, heart failure, Eisenmenger syndrome	ECG/Holter, echo, labs, clinical exam

NYHA FUNCTIONAL CLASS (ADULTS) QUICK REFERENCE

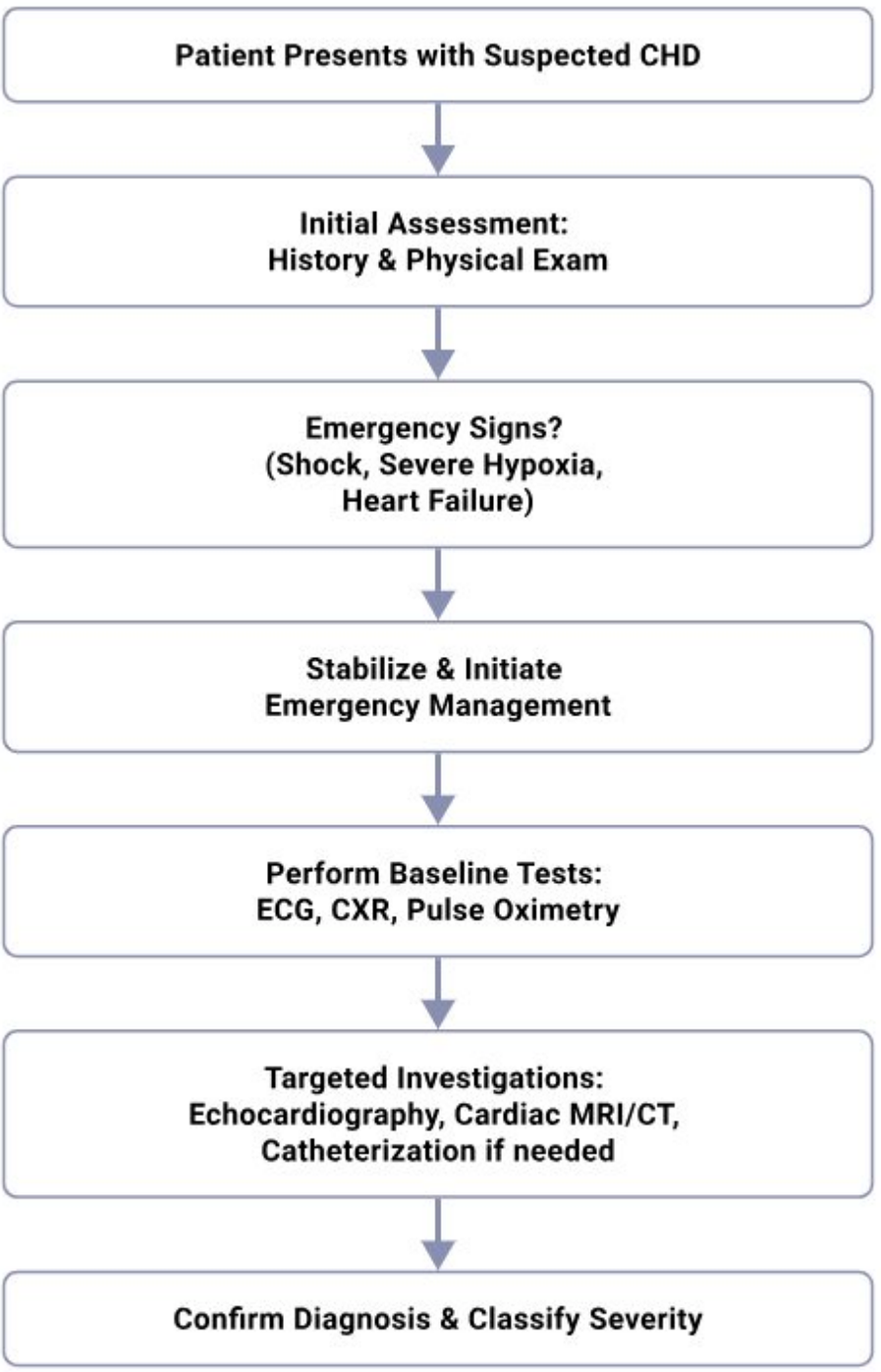
(For details see guidelines on Heart failure)

Class	Symptoms	Functional capacity
I	No symptoms with ordinary activity	No limitation
II	Dyspnea, fatigue, palpitations, or angina with ordinary activity	Slight limitation
III	Symptoms with less-than-ordinary activity (e.g., short walk, mild chores)	Marked limitation; comfortable at rest
IV	Symptoms at rest or with any activity	Unable to carry out physical activity without discomfort

Note: For infants/young children where NYHA is less applicable, use the **Ross classification**; for older children/adolescents, NYHA is commonly used alongside age-specific vital ranges and objective tests (6-minute walk, CPET).

Severity Stratification Table

Severity	Criteria	Example Conditions	Management Priority
Mild	Small defect, no pulmonary hypertension, asymptomatic	Small VSD, mild PS	Monitor, lifestyle advice
Moderate	Moderate shunt/gradient, mild symptoms	Moderate ASD, CoA	Medical therapy, consider intervention
Severe	Large shunt/gradient, pulmonary hypertension, cyanosis, heart failure	Large VSD with CHF, TOF, severe AS	Urgent referral, surgical or catheter-based intervention


Patient Presents with Suspected CHD
Initial Assessment:
History & Physical Exam
Emergency Signs?
(Shock, Severe Hypoxia,
Heart Failure)
Stabilize & Initiate
Emergency Management
Perform Baseline Tests:
ECG, CXR, Pulse Oximetry
Targeted Investigations:
Echocardiography, Cardiac MRI/CT,
Catheterization if needed
Confirm Diagnosis & Classify Severity

DIFFERENTIAL DIAGNOSIS

Certain conditions can mimic or coexist with congenital heart disease, therefore, ensuring accurate identification of the underlying lesion. It emphasizes distinguishing CHD from acquired cardiac disorders, pulmonary disease, and systemic causes of cyanosis or heart failure. A systematic approach helps avoid misdiagnosis, prevent inappropriate interventions, and ensure timely, targeted management.

A. Quick reference table of differential diagnosis

Presentation	Cardiac CHD possibilities	Important non-cardiac mimics	First tests to differentiate
Neonatal cyanosis	TGA, TOF, TAPVC, pulmonary atresia	PPHN, pneumonia, sepsis, methemoglobinemia	Pre/post-ductal SpO_2, hyperoxia test, CXR, urgent echo
Infant heart failure	Large VSD/ASD/AVSD, PDA, ALCAPA	Severe anemia, thyroid disease, myocarditis	ECG, CXR, troponin if ALCAPA, echo
Systolic murmur	Small VSD/ASD, PS/AS, BAV	Innocent murmur	Detailed auscultation, pulses/BP, echo if pathological features
Differential BP/ pulses	Coarctation, severe AS	Vasculitis, peripheral arterial disease	Four-limb BP, femoral delay, echo ± CT/MR aorta
Exertional cyanosis/dyspnea	Eisenmenger, pulmonary hypertension	Asthma, PE, ILD	Echo with RVSP, BNP, lung imaging if needed
Syncope/ arrhythmia (ACHD)	Post-repair VT/atrial flutter, AV block	Vasovagal, seizure	ECG, ambulatory monitor, electrolytes, echo
Chest pain (ACHD)	Coronary anomalies (ALCAPA, anomalous origin with inter-arterial course), ischemia in repaired CHD (post-arterial switch), aortic root dilation/ dissection in BAV/aortopathy	Musculoskeletal pain, gastro-esophageal reflux disease (GERD), anxiety/ panic	ECG, high-sensitivity troponin, echocardiography; consider coronary CTA/CMR if anomaly suspected; aortic root imaging if BAV/aortopathy

Note: When in doubt: stabilize hypoxemic or shocked patients first, obtain urgent echocardiography, and refer early to a center with pediatric cardiologist.

B. By pathophysiology to rule out (common non-cardiac causes of hypoxemia or HF mimics)

System/Pathway	Examples	Clues / First-line tests
Pulmonary causes of hypoxemia	Pneumonia, bronchiolitis, asthma, pulmonary embolism (PE), persistent pulmonary hypertension of the newborn (PPHN)	CXR; lung ultrasound; ABG; D-dimer/CTPA if PE risk; pre/post-ductal SpO_2 (PPHN)
Hematologic	Secondary polycythemia, methemoglobinemia, anemia	CBC; methemoglobin level; blood film; pulse-ox waveform vs ABG saturation gap

Metabolic/ Endocrine	Thyroid disease with cardiomyopathy or tachyarrhythmia	TSH/FT4; ECG; echo if dysfunction suspected
Systemic hypertension / renal disease	Hypertensive crisis, renal artery stenosis (mimics coarctation symptoms)	4-limb BP; urinalysis; renal ultrasound/renin profile as indicated
Valvulitis	Acute rheumatic valvulitis in adolescents vs congenital valve disease	ASO/streptococcal testing; echo for valve morphology and regurgitation
Endocarditis in ACHD	Infective endocarditis vs residual shunt/valve lesion	Blood cultures, inflammatory markers, transthoracic/transesophageal echo

Bedside differentiation of neonatal cyanosis - hyperoxia test and differential cyanosis

Hyperoxia test (how to do it)

1. Place pre-ductal probe on right hand; keep a post-ductal probe on a foot.
2. Get a baseline pre-ductal arterial blood gas (ABG) (right radial) on room air.
3. Give 100% O_2 (FiO_2 1.0) via hood/CPAP for 10 minutes.
4. Repeat pre-ductal ABG and document pre-/post-ductal SpO_2.

Cut-offs (interpretation)

- PaO_2 >150-200 mmHg or rise >100 mmHg → favors pulmonary parenchymal disease (not a fixed cardiac shunt).
- PaO_2 <150 mmHg or rise <30 mmHg despite 100% O_2 → suggests right-to-left shunt: cyanotic CHD or PPHN (persistent pulmonary hypertension of the newborn).
- PPHN clues: pre-post ductal gap (SpO_2 ≥3-5% or PaO_2 ≥10-20 mmHg higher in right hand), partial improvement with O_2/ventilation.
- Cyanotic CHD clues: minimal PaO_2 response to 100% O_2; may have single loud S2 (e.g., TGA). Murmur pattern helps (e.g., RVOT murmur in TOF).

Differential cyanosis (pre- vs post-ductal)

- Pre-ductal (right hand) > post-ductal (foot): PPHN or duct-dependent systemic flow (e.g., critical coarctation/interrupted arch) with right-to-left shunt across PDA.
- Reverse differential cyanosis (foot > right hand): Transposition of the great arteries (TGA) with PDA ± coarctation/interrupted arch.
- Treat pre/post gap ≥3-5% SpO_2 (or PaO_2 gap ≥10-20 mmHg) as significant.

Extra bedside separators

- Single loud second heart sound (S2): suggests high pulmonary pressure or great-artery malposition (e.g., TGA).
- Murmur quality: harsh ejection at LUSB (RVOT obstruction/TOF) vs often no murmur in TGA unless VSD/PS.
- Hepatomegaly: supports heart failure from volume/pressure load.

MANAGEMENT GOALS

Management of congenital heart disease aims to address both acute and long-term outcomes.

Management goals (adapt to age, lesion, severity, resources)

1. Stabilize circulation: maintain oxygen delivery and perfusion; avoid worsening shunts or raising pulmonary vascular resistance.
2. Relieve symptoms / improve function: reduce cyanosis, dyspnea, heart failure; support normal activity.
3. Prevent irreversible harm: close significant left→right shunts early to prevent pulmonary vascular disease; avoid Eisenmenger syndrome; prevent endocarditis where indicated.
4. Correct what's fixable: repair/palliate structural defects; treat anemia, infections, electrolyte issues that worsen cardiac status.
5. Limit arrhythmias and sudden death: rhythm surveillance in high-risk lesions (e.g., repaired Tetralogy of Fallot, Fontan).
6. Reduce long-term morbidity: time interventions to protect ventricular and valve function; ensure transition to adult congenital heart disease (ACHD) follow-up.
7. Maintain growth and development: nutrition support, developmental surveillance, vaccination, psychosocial care.

MANAGEMENT PRINCIPLES (STEPWISE, MULTIDISCIPLINARY)

1. Early recognition & stabilization

- Identify time-critical presentations: cyanotic spell, shock, severe heart failure.
- ABCs first. Administer Oxygen judiciously - avoid disrupting duct-dependent balance.
- Neonates with suspected duct-dependent lesions: start prostaglandin E1 promptly.

2. Precise diagnosis

- Echocardiography (2D/Doppler) to define anatomy, shunt direction, pressures, valve function.
- Classify physiology: cyanotic vs acyanotic; shunt vs obstructive. Involve a pediatric cardiologist; multiple lesions may coexist.

3. Risk stratification

- Use SpO_2, NYHA/Ross class, ventricular function, and growth.
- Flag for urgent tertiary referral: critical coarctation, transposition, obstructed TAPVR, duct-dependent systemic/pulmonary flow.

4. Multidisciplinary plan

- Coordinate cardiology, cardiac surgery, anesthesia, ICU.
- In limited-resource settings, organize teleconsults with tertiary centers.

5. Stepwise care

- Acute stabilization: secure hemodynamics and perfusion.
- Medical optimization: diuretics, afterload reduction, antiarrhythmics as indicated; targeted pulmonary vasodilators where appropriate.
- Definitive therapy: surgical repair or catheter intervention; palliation when repair must wait.
- Long-term follow-up: surveillance for residual lesions, ventricular/valve dysfunction, pulmonary hypertension, re-intervention needs.

6. Reversible factors

- Treat anemia, infections, electrolyte/acid-base problems.
- Optimize infant nutrition (calorie density, feeding support) to improve surgical readiness.

7. Emergencies to anticipate

- Atrial/ventricular arrhythmias: low threshold for cardioversion in instability.
- Systemic or single-RV circulations: high collapse risk with arrhythmia - monitor closely.
- Right-sided endocarditis: higher risk with prosthetic valves/conduits.
- Paradoxical embolism: consider in cyanotic patients with acute neurologic signs.

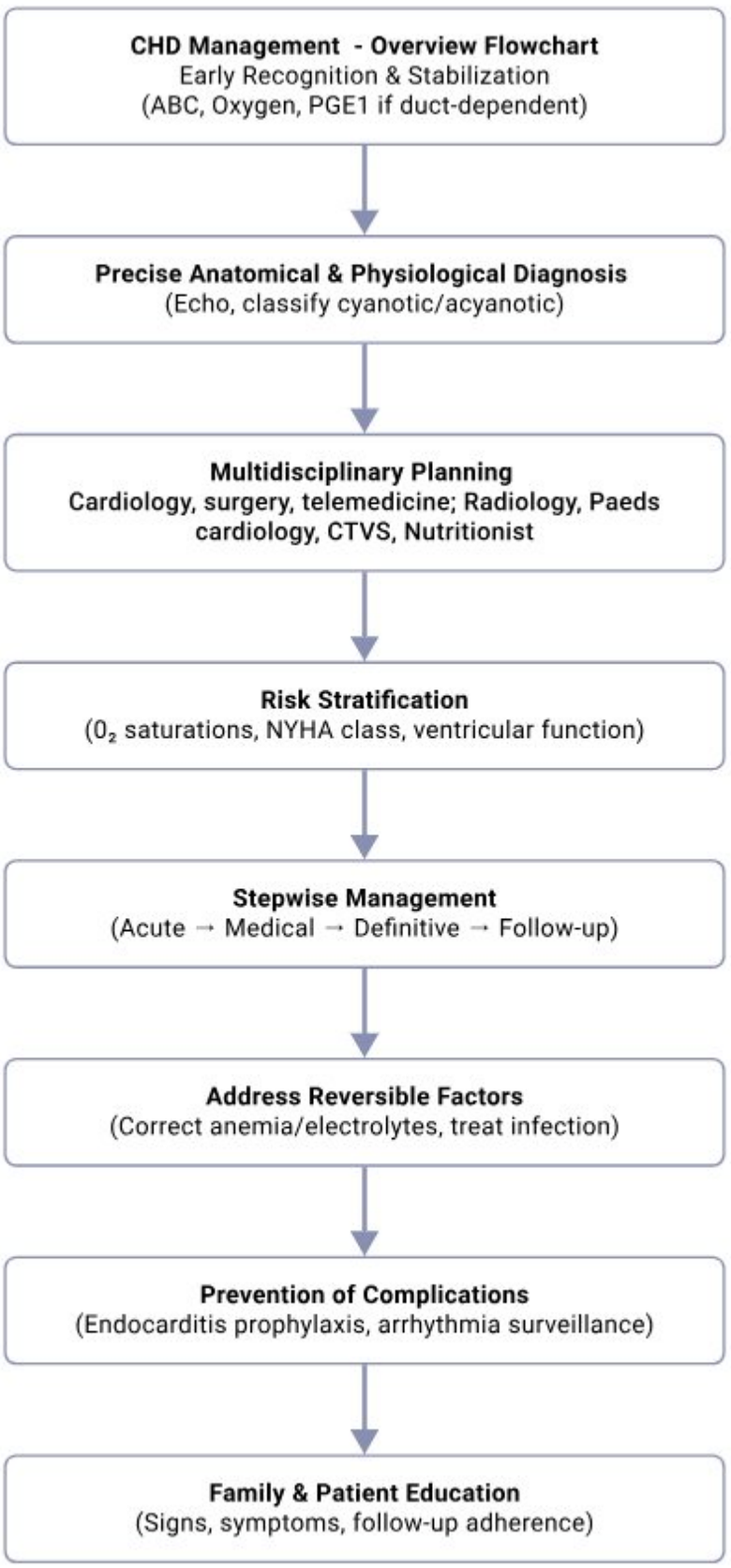
8. Prevent complications

- Endocarditis prophylaxis when indicated (prosthetic material ≤6 months or residual shunt adjacent to prosthesis, prior IE). (Details see below).
- Routine rhythm surveillance; early detection and treatment of pulmonary hypertension.

9. Shared, tiered care

- Complex CHD: shared care between local services and ACHD centers.
- Even "simple" CHD warrants periodic specialist review.
- Keep direct communication lines open between referral and local teams.

Note: These goals matter because patients with CHD are never truly "cured"; even after successful surgery, they remain at lifelong risk for complications such as arrhythmias, heart failure, or progressive valve disease. Early, goal-directed management is critical to reducing mortality, preventing avoidable hospitalizations, and supporting optimal growth and developmental outcomes. In resource-limited settings, where timely access to tertiary surgical care may be delayed, having structured, clearly defined goals helps bridge the gap by guiding stabilization, prioritizing interventions, and ensuring that patients reach definitive treatment in the best possible condition.


CHD Management - Overview Flowchart
Early Recognition & Stabilization
(ABC, Oxygen, PGE1 if duct-dependent)
Precise Anatomical & Physiological Diagnosis
(Echo, classify cyanotic/acyanotic)
Multidisciplinary Planning
Cardiology, surgery, telemedicine; Radiology, Paeds cardiology, CTVS, Nutritionist
Risk Stratification
(0_2 saturations, NYHA class, ventricular function)
Stepwise Management
(Acute → Medical → Definitive → Follow-up)
Address Reversible Factors
(Correct anemia/electrolytes, treat infection)
Prevention of Complications
(Endocarditis prophylaxis, arrhythmia surveillance)
Family & Patient Education
(Signs, symptoms, follow-up adherence)

APPROACH TO MANAGEMENT BY CHD TYPE & AGE GROUP

The approach to CHD management varies by lesion type and patient age, balancing urgent correction for critical defects in infants with long-term surveillance and complication prevention in adults with repaired or unrepaired disease. Care plans should be individualized, accounting for anatomy, physiology, prior interventions, and comorbidities.

Group	Focus	Key actions (with doses where relevant)	Monitoring & cautions
Universal first steps (any suspected critical CHD)	Stabilize, assess, confirm quickly	▪ ABC, warm, glucose; correct acidosis/ electrolytes. ▪ Pulse-ox pre/post-ductal, 4-limb BP, CXR/ ECG; Echo ASAP. ▪ Prostaglandin E_1 (alprostadil) if duct-dependent systemic or pulmonary flow possible ***0.05-0.1 µg/kg/min; titrate down to 0.01-0.025 µg/kg/min once stable; up to 0.1 µg/kg/min if needed (specialist if higher);*** Prepare for apnea and hypotension. ▪ Oxygen to target, not maximal; avoid hyperoxia if it worsens shunt balance.	▪ Continuous cardiorespiratory/ SaO_2 monitoring. ▪ Watch for PGE_1 adverse effects (apnea, hypotension, fever/flushing). ▪ Early discussion with pediatric cardiology/transport team.
Neonates	Maintain ductal patency in duct-dependent lesions; stabilize oxygen delivery	▪ Start PGE ▪ Airway readiness; secure line; start before transport. ▪ Immediate echo and surgical/catheter plan. ▪ Oxygen: target saturations per lesion; avoid hyperoxia (e.g., TGA without mixing).	▪ Anticipate apnea, hypotension, fever/flushing on PGE_1. ▪ Monitor glucose, lactate, electrolytes, urine output.
Infants & young children	Repair major shunts/ obstructions before pulmonary vascular disease	**Heart failure therapy:** ▪ Furosemide ▪ Add-on diuretics: Spironolactone/ Chlorothiazide/ Hydrochlorothiazide ▪ Afterload reduction: ACE inhibitor for LV volume load/AV valve regurgitation (dose per local protocol). ▪ Nutrition: 120-150 kcal/kg/day; fortify feeds; treat anemia.	▪ Monitor urine output, weight, $Na^+/K^+/Mg^{2+}$, creatinine. ▪ Ototoxicity risk with rapid IV furosemide/aminoglycosides. ▪ Plan timely surgical/catheter closure when indicated.
Adolescents	Residual/repaired lesions; arrhythmia and valve surveillance	▪ Rhythm assessment/management; evaluate exercise tolerance ▪ Education: infective endocarditis prophylaxis indications; sports/exercise and contraception counselling.	▪ Echo/CMR as indicated; ambulatory ECG if symptomatic. ▪ Transition planning to ACHD services.
Adults with congenital heart disease (ACHD)	Lifelong surveillance for heart failure, arrhythmias, pulmonary hypertension	▪ Regular ACHD clinic follow-up; guideline-directed HF therapy when needed. ▪ Women: pre-pregnancy risk counselling and planning. ▪ Re-interventions: valve replacements, conduit/patch issues, catheter or surgical revisions.	▪ Monitor rhythm, ventricular function, pulmonary pressures long-term. ▪ Multidisciplinary care for high-risk scenarios (cardiology, obstetrics, anesthesia).

PEDIATRIC HEART FAILURE

Symptoms by age group

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

Key signs on examination

- Tachypnea with intercostal/subcostal retractions (RR >60/min in <1 year; >50/min in 1-2 years)
- Tachycardia (HR >160/min in <1 year; >140/min in 1-2 years)
- Hepatomegaly
- Auscultation: basal crackles (limited sensitivity/specificity)
- S3 gallop, murmurs
- Raised JVP (not useful in infants)
- Peripheral edema

Red flags (Urgent escalation)

- Poor peripheral perfusion
- Oliguria/reduced urine output
- Elevated lactate
- Altered sensorium

Common heart failure mimics

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

Essential investigations

Test	Findings	Examples
Chest X-ray	Cardiac silhouette, pulmonary vasculature, PA dilation, associated skeletal abnormalities	Cardiomegaly with pulmonary plethora suggests overcirculation; oligemia suggests reduced pulmonary flow
ECG	Arrhythmia, chamber hypertrophy, ischemia; identifies some treatable causes	Deep Q waves in I, aVL, V5-V6 can suggest ALCAPA; hypocalcemia → QTc prolongation
Basic labs	Acid-base, perfusion, metabolic status	ABG, lactate, electrolytes, renal/liver function baselines and for drug monitoring
	Optional	Thyroid function test, Natriuretic Peptide (NP) Cardiac enzymes (troponin I, T, CKM) and In suspected cases of myocarditis Viral Panel
Echocardiography	Defect anatomy, ventricular function, valve regurgitation, pulmonary pressures	Obtain urgently when HF suspected; guides definitive management

Modified Ross classification for Heart failure in children

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

Age-specific normal vitals

Age group	Heart rate (awake, bpm)	Respiratory rate (/min)	Systolic BP (mmHg)	SpO_2 (room air)
Neonate (0-28 d)	100-205	40-60	~60-76	≥95%
Infant (1-12 mo)	100-180	30-53	72-104	≥95%
Toddler (1-3 y)	98-140	22-37	86-106	≥95%
Preschool (4-5 y)	80-120	20-28	89-112	≥95%
School-age (6-12 y)	75-118	18-25	97-115	≥95%
Adolescent (13-18 y)	60-100	12-20	110-131	≥95%

Use the vitals table to interpret symptom severity (tachycardia, tachypnea, hypotension) for the child's age when applying the severity criteria above.

Management goal: Correct the underlying cause; Reduce associated morbidity and mortality; Improve functional status and quality of life

Drug dose reference			
Drug	**Usual starting dose**	**Titration / max**	**Key monitoring**
Furosemide (PO/IV)	1 mg/kg/dose (neonate 0.5-1 mg/kg)	q6-12 h; max 2 mg/kg/dose; refractory: 0.05-0.2 mg/kg/h infusion	Urine output, weight, Na^+/K^+/Mg^{2+}, creatinine; ototoxicity
Spironolactone (PO)	1-3 mg/kg/day	Split q12-24 h	K^+, creatinine
Chlorothiazide (PO/IV)	5-10 mg/kg/dose q12 h	-	Na^+/K^+, volume status
Hydrochlorothiazide (PO)	1-2 mg/kg/day q12-24 h	-	Na^+/K^+, volume status
Enalapril (PO) - for LV volume load/AV valve regurgitation	0.05 mg/kg/dose once daily (or 0.1 mg/kg/day in 1-2 doses)	Titrate every 1-2 weeks to 0.1-0.4 mg/kg/day in 1-2 doses; max ~0.5-0.6 mg/kg/day (not >40 mg/day)	BP (first-dose hypotension), creatinine/eGFR, K^+; cough/angioedema. Avoid or use specialist oversight in neonates (<1 month).
Lisinopril (PO) - for LV volume load/AV valve regurgitation (school-age/adolescents)	0.07 mg/kg/day once daily (max initial 5 mg)	Titrate to 0.2-0.6 mg/kg/day once daily; max 40 mg/day	BP, creatinine/eGFR, K^+; counsel on teratogenicity (adolescents)

General Measures

- **Fluid restrictions-** in acute HF with lung congestion, peripheral edema despite diuretics and in presence of hyponatremia
- **Rest**: Restriction of activity ◦Activity as tolerated for older children with chronic compensated HF
- **Correction of Anemia** - 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 Calcium and it 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

***Note**: Always individualize by lesion, renal function, blood pressure, and local protocols.*
For details see Heart failure Guidelines

C. Level-of-Care Integration

Level	Priority Actions
Primary	Early recognition, stabilization, oxygen (if indicated), urgent referral, avoid harmful delays.
Secondary	Definitive diagnosis, stabilization, pre-surgical optimization, teleconsultation with tertiary center.
Tertiary	Complex surgery, catheter-based interventions, ACHD specialized care, advanced heart failure therapy.

General approach (all Left to Right shunts) General and Lesion-Specific Management

Goals: relieve heart failure from pulmonary overcirculation, support growth, prevent pulmonary vascular disease, and close the defect when indicated.

Table 1. General approach (applies to all left to right shunts)

Section	Key points
Initial work-up	Exam; CXR; ECG; echocardiography (defect size/type, chamber dilation, pulmonary pressures, AV valve regurgitation); ± BNP.
Medical support (when symptomatic)	▪ Diuretics (e.g., furosemide; add spironolactone if needed). ▪ Afterload reduction (ACE inhibitor) if LV volume overload/AV valve regurgitation (specialist guided). ▪ Nutrition: 120-150 kcal/kg/day; feed fortification; treat anemia. ▪ Oxygen only as needed; avoid excessive FiO_2 that worsens pulmonary overcirculation.
IE prophylaxis	Not routine for isolated ASD/VSD/PDA/AVSD. Reserve for prosthetic material within 6 months of repair, residual shunt adjacent to prosthetic material, or prior infective endocarditis.
Activity	Usually normal if small/asymptomatic. Restrict with heart failure or pulmonary hypertension.
Follow-up	Cardiology follow-up; echo interval based on lesion severity and symptoms.

Table 2. Lesion-specific management (left to right shunts)

Condition	Types / Key features	When to intervene	Method	Aftercare / Follow-up
Atrial Septal Defect (ASD)	Types: secundum (device-eligible), primum/sinus venosus/coronary sinus (surgical).	Close if any of: right-heart dilatation with Qp:Qs ≥1.5; symptoms (exercise intolerance, growth failure); paradoxical embolism. Usual timing 2-5 years; earlier if significant overload.	Transcatheter device for suitable secundum rims. Surgery for primum, sinus venosus, deficient rims, or associated PAPVR.	Aspirin low-dose ~6 months post device; IE prophylaxis 6 months after device/ surgery. Echo at 6-12 months, then as advised.
Ventricular Septal Defect (VSD)	Types: perimembranous, muscular, inlet, outlet.	Small/restrictive: observe (many close/diminish). Moderate-large (symptomatic or LV dilation): close for persistent symptoms/poor growth despite therapy, LV volume overload, recurrent respiratory infections, rising pulmonary pressures, aortic cusp prolapse/aortic regurgitation. Timing: early infancy if failure to thrive; otherwise before pulmonary vascular disease.	Surgical patch closure standard; device closure for selected muscular VSDs.	Echo follow-up; manage residual shunt or AV block if present; IE prophylaxis 6 months if prosthetic material used.
Patent Ductus Arteriosus (PDA)	Term: audible PDA with left-heart dilation, symptoms, or pulmonary overcirculation. Preterm: assess hemodynamic significance.	Close if symptomatic or with left-heart dilation/ overcirculation.	Transcatheter coil/ device preferred in term infants/children; surgery if anatomy unsuitable. Preterm: pharmacologic closure (ibuprofen/ indomethacin/ paracetamol) per protocol; ligation if refractory.	Short antiplatelet course after device (center-specific). IE prophylaxis 6 months if prosthetic material.
Atrioventricular Septal Defect (AVSD)	Common in Down syndrome; frequent AV valve regurgitation; risk of early pulmonary hypertension.	Complete AVSD: repair at 3-6 months (earlier if uncontrolled failure/PH). Partial/transitional: repair at 2-4 years if significant left AV (mitral) regurgitation or volume overload.	Medical stabilization (diuretics, specialist-guided afterload reduction), nutrition. Definitive complete repair; PA banding rarely as bridge if repair must be delayed.	Post-repair: monitor AV valve regurgitation, LV function, conduction block; manage PH if present. IE prophylaxis 6 months after prosthetic material.
Tetralogy of Fallot (TOF)	Clues: cyanosis ± spells; harsh ejection murmur at LUSB; boot-shaped heart on CXR.	Intervene per symptoms/ anatomy (See below).	Complete repair in infancy; acute spell management per separate protocol.	Standard follow-up after repair; surveillance for PR, RV dilation, arrhythmia (center-specific).

Transposition of the Great Arteries (TGA, d-TGA)	Clues: profound cyanosis day 1-2, single loud S2, minimal murmur.	Urgent stabilization in neonate with poor mixing/ restrictive atrial septum.	Stabilize: PGE_1 0.05-0.1 µg/kg/min IV; correct acidosis; Balloon atrial septostomy (BAS) if restrictive atrial septum/poor mixing. Definitive: Arterial switch (Jatene) with coronary transfer in neonatal period (ideally ≤2 weeks).	Follow-up: coronary patency, neoaortic valve regurgitation, supravalvar PS, ventricular function, rhythm.
Total Anomalous Pulmonary Venous Connection (TAPVC/TAPVR)	Clues: cyanosis, respiratory distress; CXR "snowman" in supracardiac (non-obstructed). Distinguish obstructed vs non-obstructed.	Immediate if obstructed; early even if non-obstructed.	Stabilize: oxygen/ ventilation and PEEP; treat acidosis. PGE_1 not helpful (not duct-dependent). Avoid delay for cath. Definitive: Emergency surgery if obstructed; early repair otherwise.	Follow-up: pulmonary venous stenosis, pulmonary hypertension (± inhaled NO/ sildenafil), RV function.
Coarctation of the Aorta (CoA)	Clues: weak/absent femorals, upper-lower BP gradient; shock when PDA closes.	Neonate/infant in shock needs immediate stabilization; intervene after stabilization.	Stabilize: PGE_1 0.05-0.1 µg/kg/min IV; diuretics if pulmonary edema; cautious inotropes; correct acidosis. Definitive: Surgical repair (extended end-to-end) in neonates/ infants; balloon angioplasty/stent for recoarctation or older child/adolescent.	Follow-up: recoarctation surveillance; persistent hypertension (beta-blocker/ACE-I); aneurysm surveillance with MRI/CT in older children/adults.
Single-Ventricle Physiology (e.g., HLHS, unbalanced AVSD, tricuspid atresia with PA)	Clues: duct-dependent systemic or pulmonary flow; mixing physiology; variable cyanosis.	Stage-appropriate interventions per pathway; urgent stabilization in neonate.	Stabilize: PGE_1 to maintain ductus; balance Qp:Qs - avoid hyperoxia/excessive ventilation; target SpO_2 ~75-85% unless directed; diuretics/ inotropes; correct acidosis. Staged palliation: 1. Norwood/hybrid (neonate) 2. Bidirectional Glenn (3-6 mo) 3. Fontan (2-4 y). Interstage program for weight/SpO_2/ feeds and red-flag escalation.	Follow-up: AV-valve regurgitation, arrhythmias, thromboembolism (antiplatelet/ anticoagulants per protocol), Fontan-associated issues (PLE, liver disease); exercise/cyanosis targets; vaccination/ IE prevention.

Hypercyanotic ("tet") spells management

Recognize

- Sudden deep cyanosis, irritability/crying, tachypnea, ↓ murmur intensity, possible syncope/seizures. Most common in Tetralogy of Fallot.

Immediate steps (do all promptly)

- Position: knee-chest (or squatting in older child) to raise SVR.
- Calm/sedate: minimize agitation; morphine 0.1 mg/kg IV/IM (or ketamine 0.5-1 mg/kg IV if intubating or morphine unsuitable).
- Oxygen: 100% by face mask.
- Fluids: 10-20 mL/kg normal saline bolus.
- Correct acidosis: $NaHCO_3$ 1-2 mEq/kg IV if acidotic.

Medications to break the spell

- Beta-blocker:
  - Propranolol 0.01-0.1 mg/kg IV slow push (or)
  - Esmolol bolus 100-500 mcg/kg, then 100-500 mcg/kg/min infusion.
- Alpha-agonist to raise SVR (if persistent cyanosis): Phenylephrine 5-20 mcg/kg IV bolus; repeat/titrate.
- Consider intubation with deep anesthesia (e.g., ketamine) if refractory.

If diagnosis uncertain / duct-dependent pulmonary flow

- Start PGE_1 (alprostadil) 0.05-0.1 µg/kg/min IV, prepare for apnea.

After stabilization

- Prevent recurrence: propranolol 1-2 mg/kg/day PO divided; treat iron deficiency; avoid dehydration/crying triggers.
- Definitive care: early complete repair (VSD closure + RVOT relief) 3-6 months; earlier if recurrent spells/severe RVOTO. If not immediately feasible: palliation - RVOT stent or modified BT shunt.
- Monitor/transfer: continuous SpO_2/ECG/BP; document times, doses, response; arrange urgent pediatric cardiology review.
- Follow-up focus: residual RVOTO, pulmonary regurgitation, RV dilation, arrhythmias.

When to escalate/transfer (any lesion)

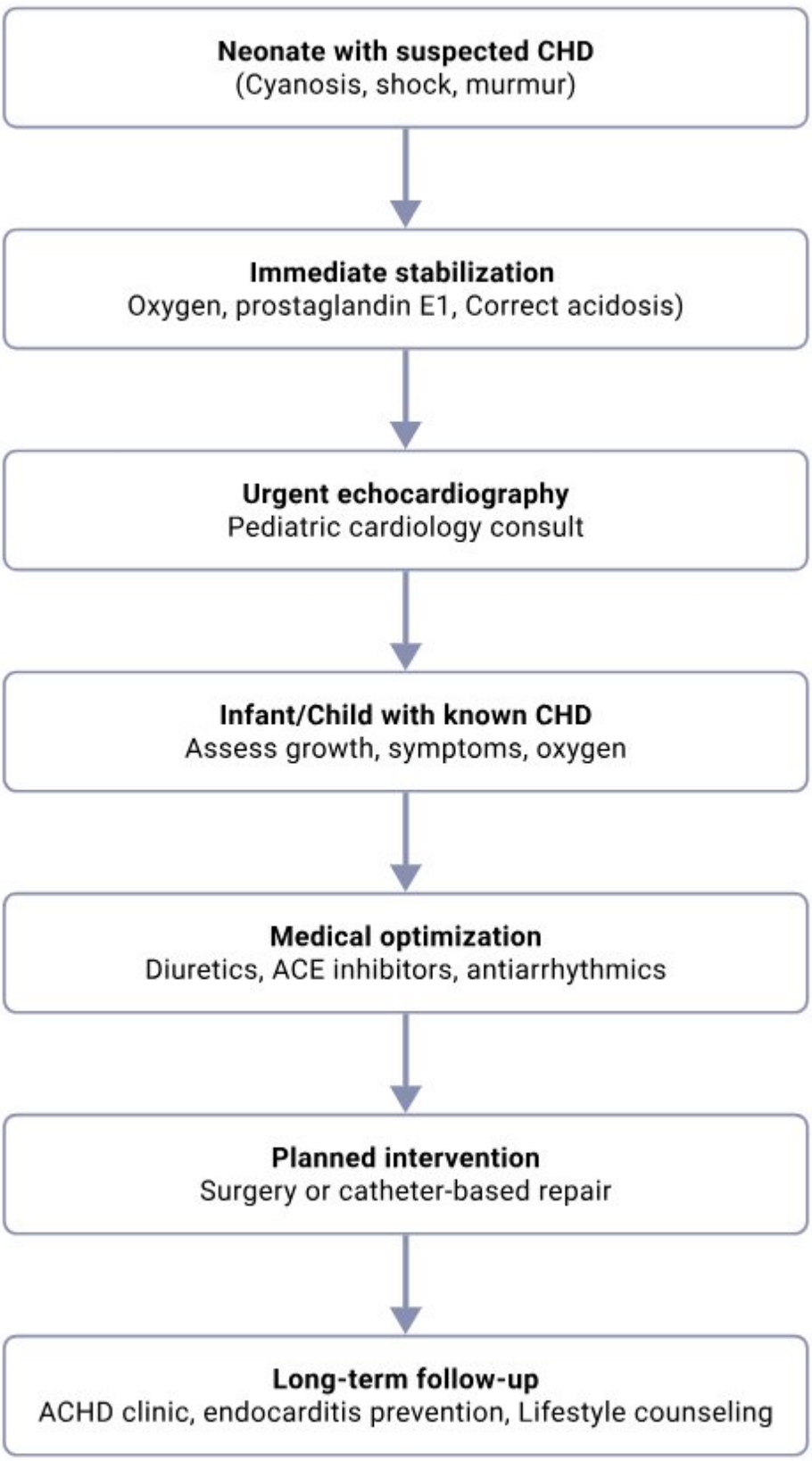
- Refractory hypoxemia/cyanotic spell, rising lactate, shock, severe heart failure, rising RVSP/PH, or cyanotic spells (critical RVOTO in ToF differential).
- Suspected duct-dependent circulation without on-site surgery - start PGE_1 and transfer now.
- Worsening pulmonary hypertension, recurrent arrhythmias, failure to thrive despite therapy.
- Any signs of Eisenmenger physiology (exercise cyanosis, differential saturation) → do not close; refer to PH/ National Cardiac Center, Pediatric Cardiologist.

Note: *Coordinate early with pediatric cardiology/CTVS. Document all pre-referral interventions (drug name-dose-route-time, oxygen modality/FiO_2, lines, response) and send with the patient.*

Indications to close the defect

Lesion	Key triggers for intervention	Usual approach
ASD (secundum)	RA/RV dilatation, Qp:Qs ≥1.5, symptoms/ paradoxical embolus	Device (if rims) or surgery
VSD	Symptoms/poor growth despite meds, LV dilation, aortic cusp prolapse/AR, rising PAP	Surgery (most); device for select muscular
PDA (term)	Symptoms, LV dilation, pulmonary overcirculation	Transcatheter device; surgery if unsuitable
AVSD (complete)	Early PH/heart failure	Complete repair at 3-6 months (earlier if needed)
AVSD (partial)	Significant L-AV valve regurgitation/volume load	Surgical repair at 2-4 y

Coordinate decisions with pediatric cardiology. Individualize by anatomy, symptoms, growth, and pulmonary pressures.


Neonate with suspected CHD
(Cyanosis, shock, murmur)
Immediate stabilization
Oxygen, prostaglandin E1, Correct acidosis)
Urgent echocardiography
Pediatric cardiology consult
Infant/Child with known CHD
Assess growth, symptoms, oxygen
Medical optimization
Diuretics, ACE inhibitors, antiarrhythmics
Planned intervention
Surgery or catheter-based repair
Long-term follow-up
ACHD clinic, endocarditis prevention, Lifestyle counseling

PHARMACOLOGICAL THERAPY

Pharmacologic management in CHD depends on the lesion type, hemodynamic status, age, and comorbidities. The primary aims are to stabilize circulation, reduce cardiac workload, control arrhythmias, manage pulmonary hypertension, and prevent complications such as heart failure or thromboembolism.

Surgical or interventional repair remains definitive for many lesions, but pharmacologic therapy bridges the period before surgery, supports postoperative recovery, and serves as long-term therapy in unrepaired or palliated defects.

Table: CHD Pharmacologic Therapy by Indication & Age

CHD / Clinical Context	Drug	Dose	Route	Duration	Cautions
Pulmonary hypertension (post-repair or unrepaired shunt)	Sildenafil	0.5-2 mg/kg/dose q6-8h (max adult 20 mg/dose)	PO	Long-term	Hypotension; avoid with nitrates
	Bosentan	2 mg/kg/dose q12h (max adult 125 mg q12h)	PO	Long-term	Monthly LFTs; teratogenic - contraception required
Atrial arrhythmias in repaired CHD	Amiodarone	Load 5-10 mg/kg IV over 30-60 min; maintain 5-15 mg/kg/day PO	IV/PO	Individualized	Monitor thyroid, liver, lungs; ECG QT
	Beta-blocker (e.g., metoprolol)	0.5-1 mg/kg/dose q6-12h (max 200 mg/day)	PO	Long-term	Bradycardia, hypotension
Cyanotic CHD with secondary polycythemia	Aspirin	3-5 mg/kg/day	PO	Long-term	GI irritation, bleeding
Endocarditis prophylaxis (high-risk CHD)	Amoxicillin	50 mg/kg (max 2 g) 30-60 min before dental procedures	PO	Single dose	If allergic: cephalexin 50 mg/kg (non-anaphylactic) or azithromycin/clarithromycin 15 mg/kg (max 500 mg). Clindamycin not recommended for dental prophylaxis
Post-op anticoagulation (prosthetic material as indicated)	Warfarin	Per INR target 2-3	PO	Long-term	INR checks; food/drug interactions
Systemic-to-pulmonary shunt patency	Aspirin	3-5 mg/kg/day	PO	Long-term	Monitor for bleeding

***Note**: Doses are usual pediatric ranges; adjust for gestational/postnatal age, renal/hepatic function, and local protocol.*

NON-PHARMACOLOGIC INTERVENTIONS

Non-pharmacological interventions in CHD focus on measures beyond drug therapy to optimize cardiovascular function, prevent complications, and improve quality of life. These include surgical or catheter-based corrections, oxygen therapy, activity modification, nutritional support, infection prevention, and structured rehabilitation programs, all tailored to the patient's age, defect type, and functional status.

Section	What to do	Who/When	Key points
Lifestyle modifications	Nutritional optimization	Infants: high-calorie feeds; fortified breast milk/formula for failure to thrive. Children/adolescents: balanced diet to support growth, prevent obesity. Adults: low-salt, heart-healthy diet for comorbidities.	Monitor weight/centiles; treat anemia; dietitian input before surgery.
	Physical activity (tailored)	Base on defect severity and repair status.	Avoid high-intensity competitive sports in unrepaired cyanotic lesions or severe pulmonary hypertension; encourage daily moderate activity otherwise.
	Weight management	All ages.	Reduces HF symptoms and peri-operative risk.
Infection prevention	Endocarditis prophylaxis	High-risk CHD: prosthetic valves/ material, unrepaired cyanotic CHD, repaired CHD with prosthetic material ≤6 months or residual defect.	Use dental prophylaxis per guideline; emphasize oral hygiene.
	Vaccination	Age/season appropriate.	Influenza, pneumococcal, COVID-19, and respiratory syncytial virus (RSV) if available.
	Dental care	Regular check-ups/cleaning.	Lowers bacteremia risk.
Cardiac rehabilitation	Structured exercise + education after repair.	Post-operative or deconditioned patients.	Improves functional capacity and quality of life; customize intensity.
Device support	Pacemaker	Bradyarrhythmias or conduction disease after repair.	Choose device/lead strategy with electrophysiology team.
	Implantable cardioverter-defibrillator (ICD)	High-risk ventricular arrhythmias or secondary prevention.	Program to lesion-specific risks; counsel on activity and shocks.
Psychosocial support	Counseling, school/work support, transition planning.	Adolescents moving to adult care; families under chronic stress.	Screen for anxiety/depression; connect to social services.
Telemedicine & remote monitoring	Virtual reviews, symptom/ weight logs, oximetry where useful.	Resource-limited or geographically dispersed settings (e.g., Maldives).	Enables early detection of deterioration and faster intervention.

ASSESSMENT OF RESPONSE

The assessment of response in CHD management involves systematically evaluating clinical improvement, hemodynamic stability, and resolution or reduction of symptoms following treatment. This helps determine treatment effectiveness, guides further intervention, and ensures timely detection of complications or treatment failure.

Key Parameters to Monitor

Domain	Improvement / Stable Indicators	Red Flags / Worsening Signs	Assessment Frequency
Clinical symptoms	Better exercise tolerance/feeding/ growth; less dyspnea, fatigue, palpitations, cyanosis; no syncope, chest pain, or edema	Sudden drop in exercise/feeding tolerance; worsening dyspnea, orthopnea, PND; new chest pain, fainting; rapid weight gain/edema	Daily caregiver/patient logs; review every 2-4 weeks during titration, then every 3-6 months when stable
Physical exam	Stable/improved heart sounds and murmurs; no gallop; no ↑ hepatomegaly/JVD; warm extremities, good perfusion	New S3; rising hepatomegaly/JVD; cold extremities/poor perfusion; rapidly increasing edema	Each visit; weekly-biweekly if high-risk/ unstable
Vital signs	Targets: HR within age range; SBP ≥ hypotension cut-off (see table below); BP not persistently >95th centile; SpO_2 ≥95% on room air (or per lesion plan in cyanotic CHD); weight trending appropriately	Persistent tachycardia or bradycardia for age; SBP < cut-off or uncontrolled hypertension; SpO_2 <90% or >3% drop from baseline (cyanotic CHD); fever with decompensation	Daily home HR/BP/ weight where feasible; check at every clinic visit
Objective tests	ECG: no new arrhythmias; Echo: better EF, less regurg/stenosis, stable anatomy; BNP/NT-proBNP falling	New/worsening arrhythmias; Echo: ↓ EF, ↑ valve disease, chamber dilation; BNP/NT-proBNP rising	ECG + labs q3-6 months; Echo q6-12 months or earlier if symptomatic
Functional capacity	Age-appropriate milestones; improved NYHA/Ross class; better 6-min walk (adolescents/adults)	Milestone regression; worse NYHA/ Ross class; reduced 6-min walk/ early fatigue	Each visit (2-4 weekly during adjustments; 3-6 monthly when stable); functional tests annually

Age-specific vitals

Age group	Heart rate (bpm)	Respiratory rate (/min)	Systolic BP (mmHg)	Hypotension cut-off (PALS)	SpO_2 target
Neonate (0-28 d)	100-205	40-60	~60-76	<60	≥95% (unless cyanotic plan)
Infant (1-12 mo)	100-180	30-53	72-104	<70	≥95%
Toddler (1-3 y)	98-140	22-37	86-106	<70 + 2×age	≥95%
Preschool (4-5 y)	80-120	20–28	89–112	<70 + 2×age	≥95%
School-age (6-12 y)	75-118	18–25	97–115	<70 + 2×age	≥95%
Adolescent (13-18 y)	60-100	12–20	110–131	<90	≥95%

Notes:

- Hypertension in children is BP ≥95th percentile for age/sex/height (use charts); treat persistent elevations or end-organ signs.
- Cyanotic CHD may have individualized SpO_2 targets- use the cardiology plan; a >3% fall from baseline is concerning.
- Use trends (vitals + weight) rather than single values to judge stability.

FOLLOW-UP

The purpose of follow-up in CHD is to maintain symptom control, detect late complications such as arrhythmias, valve dysfunction, ventricular failure, or residual shunts at an early stage, and adjust treatment based on the patient's growth, age, and disease progression.

Follow-Up Strategy by Stability Status

Criteria Type	Indicators	Action
Step-Up (Escalation of Care)	▪ Persistent/worsening cyanosis, dyspnea, fatigue, or edema despite optimized therapy ▪ New murmur or change in character of existing murmur ▪ Worsening ventricular function on echocardiography ▪ New-onset arrhythmias ▪ Unexplained drop in oxygen saturation ▪ Post-surgical complications (e.g., wound infection, pericardial effusion, thrombosis)	Urgent referral to secondary or tertiary care for specialized evaluation and intervention *referral to where cardiology services are available. Pediatric cardiology where applicable
Step-Down (De-escalation of Care)	▪ Stable hemodynamics for at least 6 months ▪ No new symptoms or signs of deterioration ▪ Stable postoperative anatomy and function ▪ Maintenance on minimal effective medical therapy ▪ Good functional status for age	Continue periodic follow-up with longer intervals (routine outpatient monitoring)

Follow-up post repair

Patient Category	Follow-Up Frequency	Follow-Up Components
Stable post-repair without complications	Every 6–12 months	**History**: Exercise tolerance, feeding, growth, activity limitation **Exam**: Murmurs, heart sounds, HF signs, peripheral perfusion **Investigations**: ECG, echocardiogram, chest X-ray as indicated **Medication Review**: Adjust for weight/age, adherence check **Lifestyle**: Activity restrictions, diet, infection prophylaxis **Education**: Warning signs, adherence, follow-up importance
Stable with mild residual lesions	Every 3–6 months	Same as above, with closer surveillance of residual lesions
Unrepaired moderate/ severe CHD awaiting surgery	Every 1–3 months	Same as above, with emphasis on surgical planning and deterioration risk
Complex CHD or post-palliation	Monthly to 3-monthly	Same as above, with detailed monitoring of complications and growth
Symptomatic deterioration	Immediate review	Full reassessment with urgent investigations, stabilization, and surgical/interventional referral if needed

PROGNOSIS

- The prognosis of CHD varies widely depending on the type of defect, severity at diagnosis, timing and success of intervention, and presence of comorbidities.
- Survival is now high: >90% of children with congenital heart disease reach adulthood, including many complex cases.
- Not "cured": late complications are common (arrhythmias, heart failure, pulmonary hypertension, residual/recurrent lesions) often decades after repair.
- Mild lesions (small ASD/VSD): excellent outlook; many close spontaneously or need just one procedure.
- Moderate lesions (repaired Tetralogy of Fallot, coarctation): good quality of life, but lifelong surveillance is essential.
- Severe lesions (single ventricle physiology, transposition): higher morbidity and mortality despite palliation due to complex anatomy/physiology.
- Resource-limited settings: delays in diagnosis, limited surgical access, and weak follow-up worsen outcomes; early detection, timely referral, structured follow-up, and caregiver education are the levers that improve survival.

Prognosis Stratification Table by CHD Type

CHD Type	Prognosis	Key Predictors of Outcome
Simple CHD (e.g., ASD, small VSD, PDA)	Excellent with timely intervention; near-normal life expectancy	Early diagnosis and repair, absence of pulmonary hypertension, good ventricular function
Moderate CHD (e.g., repaired TOF, coarctation of aorta, Ebstein anomaly)	Good but may require lifelong follow-up and re-interventions	Ventricular function, residual defects, arrhythmia burden, pulmonary pressures
Severe CHD (e.g., univentricular heart, Eisenmenger syndrome, transposition of great arteries)	Variable; reduced life expectancy; high morbidity	Surgical palliation success, oxygen saturation, functional status, complications
CHD with Pulmonary Hypertension	Poor if irreversible; better if reversible with intervention	Severity and reversibility of PH, timing of repair, response to therapy
CHD with Heart Failure	Guarded prognosis; depends on HF management and comorbidities	Ejection fraction, NYHA class, renal function, adherence to treatment

REFERRAL PATHWAYS

Referral pathways for CHD must ensure timely access to specialized care, especially in resource-limited settings. The approach should be tiered according to the urgency of the patient's condition and the level of healthcare facility.

Referral category	Indicators	Actions / stabilization	Level of care	Documentation - pre-referral (required)
Emergency	Severe cyanosis unresponsive to care; shock/severe HF; refractory arrhythmias; acute post-op decompensation; critical CHD at birth (e.g., TGA, HLHS)	Oxygen; PGE1 for duct-dependent lesions; arrhythmia control; immediate transfer	Tertiary (peds/ adult cardiology + surgery)	Time-stamped vitals; O_2 modality/FiO_2/flow; PGE1 dose/rate/time; antiarrhythmic given (drug/ dose/route/time); IV/IO lines and fluids; ABG/ECG/CXR/ echo summary; response to therapy; adverse events; accepting center/consult name and time; transport mode/escort level
Urgent (24–72 h)	New moderate–severe CHD with symptoms; persistent cyanosis; pulmonary hypertension signs; worsening exercise tolerance; progressive valve disease	Diuretics; oxygen as indicated; rapid specialist evaluation	Secondary/ Tertiary	Vitals trend; drugs given (name/dose/route/time: furosemide, O_2); labs/ECG/ echo summary; growth/ feeding data; risk flags; referral request time and plan
*Routine	Stable repaired CHD; mild valve/shunt; controlled arrhythmias	Regular follow-up; elective specialist review	Secondary/ Tertiary (OPD)	Last clinic summary; meds list; recent ECG/echo numbers; issues to address at next level; next appointment/referral date

***Policy note**: Coverage changes; always confirm current **Asandha** rules before referral and brief the family on expected costs.*

Note 2: A structured referral form improves clarity and reduces delays to include complete medical history, previous surgical/interventional details, recent imaging, and treatment given en route.

Tiered Referral Approach by Level of Care

- **Primary Care**: Initial recognition, basic stabilization, ECG and chest X-ray, oxygen therapy if needed, urgent communication with referral centre.
- **Secondary Care**: Echocardiography, initiation of pharmacologic therapy, risk stratification, preparation for transfer.
- **Tertiary Care**: Advanced diagnostics, surgical/interventional procedures, complex arrhythmia management, advanced heart failure therapy.

Adaptations for Maldives & Southeast Asia.

In regions with limited tertiary care access, referral pathways should integrate:

- Telecardiology consultations for rapid expert input
- Scheduled outreach cardiac clinics for stable patients
- Use of air/sea ambulance for critical transfers
- Regional coordination between islands/districts and national cardiac centres

COMPLICATIONS

Complications in CHD can arise from the native cardiac defect, its physiological consequences, surgical or catheter-based interventions, and long-term sequelae of residual lesions. These can present in both pediatric and adult populations, though their timing, severity, and impact differ by defect type, repair status, and patient comorbidities.

Category	Examples	Key Notes
Immediate Complications (perioperative/early)	▪ Severe HF from large L→R shunts ▪ Persistent cyanosis in duct-dependent lesions ▪ Arrhythmias post-surgery ▪ Low cardiac output syndrome ▪ Bleeding, infection ▪ Post-catheter: vascular injury, device migration, pericardial effusion	Related to acute hemodynamic effects of the defect or intervention; require early recognition and management
Late Complications	▪ Arrhythmias (AF, flutter, VT) ▪ Progressive ventricular dysfunction ▪ Pulmonary hypertension ▪ Residual shunts ▪ Valve regurgitation/stenosis	Can develop years/decades after initial repair; long-term follow-up essential
Cyanotic CHD Complications	▪ Secondary erythrocytosis, hyperviscosity ▪ Coagulation disorders, stroke ▪ Paradoxical embolism ▪ Brain abscess	Due to chronic hypoxemia and right-to-left shunting
Acyanotic CHD Complications	▪ Eisenmenger syndrome → irreversible pulmonary vascular disease	Contraindication to surgical repair once developed
Syndrome-Specific (e.g., Fontan circulation)	▪ Protein-losing enteropathy ▪ Plastic bronchitis ▪ Liver disease ▪ Heart failure	Unique to palliated lesions with single-ventricle physiology
Infective Endocarditis	▪ Higher risk with prosthetic material, unrepaired cyanotic CHD, residual lesions	Emphasize prevention: oral hygiene, prophylaxis in high-risk cases
Neurodevelopmental Complications	▪ Cognitive delays ▪ Motor impairments ▪ Behavioral disorders	Especially in children post-bypass or with complex CHD; require early screening and intervention
Adult CHD-Related Complications	▪ **Reproductive**: high maternal/fetal risk in pregnancy with complex lesions ▪ **Psychosocial**: employment limitations, reduced QoL	Long-term multidisciplinary care needed

Early recognition and proactive surveillance of complications are vital. This requires structured follow-up in specialized CHD clinics, adherence to surveillance protocols (including echocardiography, cardiac MRI, and exercise testing), and timely escalation of care. In low-resource settings, developing streamlined referral pathways and leveraging telemedicine can mitigate the risk of late presentation and poor outcomes.

PREVENTION AND HEALTH PROMOTION

Prevention and health promotion in CHD span the spectrum from preconception care to lifelong follow-up. Multisectoral approach with strong maternal and antenatal care, newborn screening, pediatric cardiology, and adult CHD clinics supported by solid data systems and clear national policy can significantly improve survival, stronger quality of life, and improved long-term outcomes for people with CHD.

Level / Area	What to do	Who / When	Implementation notes
Primary prevention (maternal health)	Adequate antenatal care; maternal vaccination (e.g., rubella); folic acid; strict control of diabetes and phenylketonuria; avoid teratogenic drugs, alcohol, tobacco, recreational drugs; rubella immunity screening; Zika precautions where relevant	All women of reproductive age; preconception and during pregnancy	Use checklists at first ANC; pharmacy alerts for teratogens; dietitian input for diabetes/PKU control
Antenatal/birth surveillance	Second-trimester anomaly scan with 4-chamber and outflow tract views; if risk factors (maternal diabetes, family history), abnormal screening, or genetic/extracardiac anomalies → fetal echocardiogram at 18–22 weeks	Pregnant persons at 18–22 weeks; earlier if indicated	Standardize scan protocol; fast-track fetal echo referral; plan perinatal delivery at equipped centers
Public awareness	Promote early antenatal registration and healthy maternal lifestyle	Community level, primary care	Mass media + community health workers; simple posters/checklists
Secondary prevention (early detection)	Universal newborn pulse-oximetry; targeted echo for at-risk infants; careful cardiac auscultation and growth monitoring at well-child visits	Birth facilities; routine pediatric visits	Train district staff; maintain pulse-ox logs; referral triggers and transport pathways
Tertiary prevention (minimize disability)	Structured follow-up in CHD clinics; timely surgery/catheter interventions; infective endocarditis prophylaxis when indicated; lifelong surveillance for ACHD (arrhythmias, PH, ventricular dysfunction)	Pediatric and adult CHD services	Use recall systems; transition clinics for adolescents; standardized post-op protocols
Health-system integration	Embed CHD within Maldives NCD Plan; tiered care (primary→secondary→tertiary); expand tele-cardiology; ensure access to echocardiography	National and regional planners	Define referral SLAs; equipment and maintenance plans; teleconsult hours and hubs
School/ community programs	Education on heart-healthy behaviors; adherence to follow-up; recognition of warning signs needing urgent care	Schools, community outreach	Include BP, BMI, fitness modules; caregiver teaching aids
Data & policy (cross-cutting)	Robust health information systems, dashboards, and national policy alignment	MoH/partners	Track screening coverage, time-to-diagnosis, surgical wait times, outcomes

PATIENT EDUCATION

Objectives: Patient education in CHD is essential to ensure adherence to treatment, timely recognition of complications, and long-term quality of life.

- Education should begin at the time of diagnosis and continue through all stages of care, adapted to the patient's age, literacy level, and cultural context.
- Patients and caregivers should have a clear understanding of the specific CHD diagnosis, its potential complications, and the expected course of management.
- They must be able to recognize early warning signs of deterioration such as increasing breathlessness, cyanosis, reduced exercise tolerance, palpitations, syncope, or new swelling of the legs or abdomen. The importance of strict adherence to prescribed medications, including dosing schedules and potential side effects, should be emphasized.
- Patients should understand the need for regular follow-up visits, periodic imaging, and laboratory assessments to monitor disease progression and detect late sequelae.
- Education should also cover lifestyle modifications, including avoiding tobacco and excessive alcohol, maintaining an appropriate diet, engaging in safe physical activity as advised by the treating team, and preventing infections through vaccination and prophylaxis where indicated.

Instructions to Patient/Caregiver

Patients and caregivers should be provided with clear, practical, and actionable instructions to support day-to-day management of CHD These instructions should be reinforced verbally, in writing, and, where possible, through visual aids to ensure understanding. Key guidance includes:

Instructions to patient/caregiver

Topic	Do	Don't	Why / When to call for help
Medicines	Take exactly as prescribed; keep an updated med list; bring it to every visit.	Don't skip doses or adjust on your own.	Wrong dosing causes decompensation. Call if doses are missed repeatedly or side effects occur (vomiting, dizziness, rash, very slow/fast pulse).
Follow-up	Attend all appointments; if you can't, rebook promptly.	Don't assume "I feel fine" means no visit needed.	CHD can worsen silently. Call if you've missed a visit or new symptoms appear.
Warning signs	Watch for SOB, blue lips/fingers, fainting, chest pain, fast/irregular heartbeat, swollen legs/abdomen, reduced activity/feeds.	Don't wait it out if symptoms escalate.	Urgent care now if severe breathlessness, fainting, chest pain, SpO_2 <90% (if you monitor), or rapid swelling/weight gain.

Vaccines & infection	Keep influenza, pneumococcal, COVID-19 (and RSV if eligible) up to date; daily dental hygiene; seek care for fever.	Don't ignore dental problems or untreated infections.	Prevents pneumonia and infective endocarditis (IE). Call if fever >38°C persists or dental infection develops.
IE prophylaxis	Tell dentists and clinicians about CHD; take pre-dental antibiotic if you're in a high-risk group.	Don't take antibiotics "just in case" if not indicated.	Cuts IE risk. Ask your team if you qualify.
Activity	Follow individual activity plan; stay hydrated; warm up and pace.	Don't do high-intensity/competitive sports with unrepaired cyanotic lesions or severe pulmonary hypertension.	Overexertion can be dangerous. Call if exercise tolerance drops suddenly.
Diet & weight	Use a heart-healthy, low-salt diet; infants may need fortified feeds; track weight/centiles.	Don't use crash diets or high-salt processed foods.	Supports growth and reduces heart failure symptoms. Call dietitian if weight is falling or rising fast.
Procedures & surgery	Carry a medical alert card/bracelet; tell every provider (including dentist) about your CHD and medicines.	Don't undergo procedures without declaring CHD/ anticoagulants.	Prevents anesthesia/bleeding mishaps. Call your team for pre-op clearance.
Records	Keep a CHD care plan, recent echo/ ECG summaries, and emergency contacts together.	Don't rely on memory during emergencies.	Speeds safe care during transfers.
Local support (Maldives)	Ask for visual aids, family-centered teaching, and community health worker support; use tele-follow-up where offered.	Don't skip education because of language/literacy barriers - ask for tailored materials.	Improves understanding and adherence across islands.

Emergency numbers and where to go: write them here → _____.
Diagnosis & key meds: _____. **Next review date:** _____.

REFERENCES

1. Marelli AJ, Ionescu-Ittu R, Mackie AS, et al. Lifetime prevalence of congenital heart disease in the general population (2000–2010). Circulation. 2014;130(9):749-756.
2. van der Linde D, Konings EE, Slager MA, et al. Birth prevalence of congenital heart disease: systematic review and meta-analysis. J Am Coll Cardiol. 2011;58(21):2241-2247.
3. Hoffman JI, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol. 2002;39(12):1890-1900.
4. Brida M, Diller GP, Gatzoulis MA. Interventions for adults with CHD: Part I-operations. Circulation. 2018;138(14):1426-1436.
5. Dellborg M, et al. Adult congenital heart disease (survival/outcomes review). Circulation. 2024. https://www.ahajournals.org/doi/10.1161/CIRCULATIONAHA.123.066983
6. Cotts TB, Budts W, Prokšelj K, et al. Adults with congenital heart disease: essentials for cardiologists. Eur Heart J. 2024;45:4783-4796.
7. Warnes CA, Liberthson R, Danielson GK, et al. Changing profile of CHD in adult life (historical). J Am Coll Cardiol. 2001;37(5):1170-1175.
8. Erica Sood, Jane, W. Newburger, Julia S. Anixt, Adam R. Cassidy, Jamie L. Jackson, Richard A. Jonas, et al on behalf of the American Heart Association Council on Lifelong Congenital Heart Disease and Heart Health in the Young and the Council on Cardiovascular and Stroke Nursing. Neurodevelopmental Outcomes for Individuals with Congenital Heart Disease: Updates in Neuroprotection, Risk-Stratification, Evaluation, and Management: A Scientific Statement from the American Heart Association. Circulation 2024. Volume 149, Number 13. https://doi.org/10.1161/CIR.00000000000012
9. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for management of adults with congenital heart disease. J Am Coll Cardiol. 2019;73(12):e81-e192.
10. Baumgartner H, De Backer J, Babu-Narayan SV, et al. 2020 ESC guidelines for adult congenital heart disease. Eur Heart J. 2021;42(6):563-645.
11. Saxena A, Relan J, Agarwal R, et al. Indian timing/indication guidelines for common CHD. Ann Pediatr Cardiol. 2019;12(3):254-286.
12. Saxena A, Relan J, Agarwal R, Awasthy N, Azad S, Chakrabarty M, et al. Guidelines for the management of common congenital heart diseases in India: A consensus statement on indications and timing of intervention. Indian Heart J. 2019 May-Jun;71(3):207-223. doi: 10.1016/j.ihj.2019.07.006. Epub 2019 Aug 12. PMID: 31543193; PMCID: PMC6796629.
13. IAP Specialty Series on Pediatric Cardiology. R. Krishna Kumar, Shakuntala S. Prabhu, Shreepal Jain, Sumitra Venkatesh, M. Zulfikar Ahamed (eds); Jaypee Brothers Medical Publishers 2021. ISBN: 9789390595143

14. Indian Guidelines for Indications and Timing of Intervention for Common Congenital Heart Diseases. Indian Pediatrics, 2020;57:143–157

15. Indian Academy of Pediatrics, Cardiology Chapter. Consensus statement on management of congenital heart disease. Indian Pediatr. 2019;56(8):645-653.

16. ESC Working Group on Adult Congenital Heart Disease. Transition to adulthood: position paper. 2021.

17. Guidelines for the Management of Congenital Heart Diseases in Childhood and Adolescence. Cardiol Young. 2017 Jun;27(S3):S1-S105. doi: 10.1017/S1047951116001955. PMID: 28972464.

18. Plana MN, Zamora J, Suresh G, et al. Pulse-oximetry screening for critical CHD. Cochrane Database Syst Rev. 2018;3:CD011912.

19. CDC. Congenital Heart Defects. https://www.cdc.gov/heart-defects/index.html

20. American Academy of Pediatrics. Newborn screening for critical congenital heart defect (CCHD). https://www.aap.org/en/patient-care/congenital-heart-defects/newborn-screening-for-critical-congenital-heart-defect-cchd/

21. Health Protection Agency, Maldives. National Protocol for Screening of Critical Congenital Heart Diseases of Newborns with Pulse Oximetry. 2023. https://hpa.gov.mv/wp-content/uploads/2023/04/Pulse-Oximetry-Screening-for-Critical-Congenital-Heart-Disease-.pdf

22. Franklin RC, Jacobs JP, Krogmann ON, et al. Nomenclature and the International Pediatric and Congenital Cardiac Code. Cardiol Young. 2008;18(S2):70-80.

23. Surkova E, Brida M, Muraru D, et al. Three-dimensional echocardiography in adults with CHD: ESC/EACVI statement. Eur Heart J Cardiovasc Imaging. 2025;26(7):1244-1269.

24. Moons P, Luyckx K, Thomet C, et al. Patient-reported outcomes in adults with CHD: recommendations. Int J Cardiol. 2021;331:44-50.

25. World Health Organization. Birth defects surveillance: a manual for programme managers. Geneva: WHO; 2020.

26. Ministry of Health, Maldives. National Action Plan for the Prevention and Control of Noncommunicable Diseases 2021–2030. Malé: MoH; 2021.

27. Zühlke L, Engel ME, Karthikeyan G, et al. The REMEDY study. Eur Heart J. 2015;36(18):1115-1122.