SPECIAL ARTICLE

# Indian guidelines for indications and timing of intervention for common congenital heart diseases: Revised and updated consensus statement of the Working group on management of congenital heart diseases

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## ABSTRACT

A number of guidelines are available for the management of congenital heart diseases (CHD) from infancy to adult life. However, these guidelines are for patients living in high-income countries. Separate guidelines, applicable to Indian children, are required when recommending an intervention for CHD, as often these patients present late in the course of the disease and may have coexisting morbidities and malnutrition. Guidelines emerged following expert deliberations at the National Consensus Meeting on Management of Congenital Heart Diseases in India, held on August 10 and 11, 2018, at the All India Institute of Medical Sciences. The meeting was supported by Children's HeartLink, a nongovernmental organization based in Minnesota, USA. The aim of the study was to frame evidence-based guidelines for (i) indications and optimal timing of intervention in common CHD; (ii) follow-up protocols for patients who have undergone cardiac surgery/catheter interventions for CHD; and (iii) indications for use of pacemakers in children. Evidence-based recommendations are provided for indications and timing of intervention in common CHD, including left-to-right shunts (atrial septal defect, ventricular septal defect, atrioventricular septal defect, patent ductus arteriosus, and others), obstructive lesions (pulmonary stenosis, aortic stenosis, and coarctation of aorta), and cyanotic CHD (tetralogy of Fallot, transposition of great arteries, univentricular hearts, total anomalous pulmonary venous connection,

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Ebstein's anomaly, and others). In addition, protocols for follow-up of postsurgical patients are also described, disease wise. Guidelines are also given on indications for implantation of permanent pacemakers in children.

**Keywords:** Congenital heart disease, intervention, surgery

## INTRODUCTION

Congenital heart diseases (CHDs) are the most common birth defects, responsible for nearly one-third of all congenital birth defects.[1] The birth prevalence of CHD is reported to be 8–12/1000 live births.[2,3] One-fifth of these babies have critical heart disease requiring very early intervention. Advances in pediatric cardiology and cardiac surgery have made it possible to repair or palliate most of the CHDs including the complex ones. If access to screening, early diagnosis, and treatment is available, over 90% of patients born with CHD survive to adult life with good long-term outcome.[4] Most middle- and low-income countries lack such advanced level of care for children with CHD. Considering a birth prevalence as 9/1000, the estimated number of children born with CHD every year in India approximates 240,000, posing a tremendous challenge for the families, society, and health-care system. Approximately 10% of infant mortality in India may be accounted for, by CHDs.

## JUSTIFICATION FOR DEVELOPING INDIAN GUIDELINES

Evidence-based recommendations for the management of CHD have been published by task force members from a number of national and international associations, but these are primarily meant for children born in high-income countries. Applicability of these guidelines to the Indian population with CHD is likely to be limited. Majority of patients with CHD are not diagnosed in antenatal period and often present late in the course of the disease. These patients are often underweight, malnourished, and have comorbidities such as recurrent infections and anemia. Many of late presenters have advanced level of pulmonary hypertension, ventricular dysfunction, hypoxia, polycythemia, etc. The outcome after surgery in such patients is expected to be suboptimal with long periods of mechanical ventilation and stay in intensive care. Modifications in the treatment protocol may be required for optimizing the outcomes. All these factors justify the need for separate guidelines for the management of CHD in India, including the timing of intervention.

A statement on "consensus on timing of intervention for common congenital heart disease" which originated from the Meeting of Working Group on Management of Congenital Heart Disease in India, was published in 2008.[5] This statement was revised and updated in the subsequent National Consensus Meeting, which was held in New Delhi after a gap of 10 years, in August 2018. In the intervening 10 years, a number of pediatric cardiac centers have been established, and overall, the numbers of interventions have increased by several folds. Considering the growing population of postoperative patients including those needing regular follow-up, we added guidelines and protocols for follow-up of these patients.

## PREAMBLE

1. Every pediatrician/cardiologist/other health-care provider must strive to get a complete diagnosis on a child suspected of having heart disease, with the help of a higher center, if needed.
2. The proposed guidelines are meant to assist the health-care providers (pediatrician, cardiologist, and pediatric cardiologist) for managing cases with CHD in their practice. While these may be applicable to the majority, each case needs individualized care, and exceptions may have to be made. Guidelines are intended to define practices, meeting the needs of patients in most, but not all, circumstances and should not replace clinical judgment.
3. These guidelines are in reference to the current health-care scenario prevalent in India. Subsequent modifications may be necessary in future as the pediatric cardiology practice evolves.
4. The recommendations are classified into three categories according to their strength of agreement:
   - Class I: Is recommended/is indicated. General agreement that the given treatment or procedure is beneficial, useful, and effective.
   - Class II: Conflicting evidence and/or a divergence of opinion or both about the usefulness/efficacy of the given treatment or procedure.
     - IIa: Should be considered. Weight of evidence/opinion is in favor of usefulness/efficacy.
     - IIb: May be considered. Usefulness/efficacy is less well established.
   - Class III: Is not recommended. Evidence or general agreement that the given treatment or procedure is not useful/effective and in some cases may be harmful.

### Aims and objectives

1. To outline the optimal timing of intervention in common CHDs.
2. To formulate the guidelines and protocols for follow-up of patients who have undergone surgery/catheter interventions for CHD.

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3. To formulate the guidelines for the use of pacemakers in children.

# GUIDELINES FOR INDIVIDUAL CONGENITAL HEART DEFECTS

**ATRIAL SEPTAL DEFECT**

*Background*
Atrial septal defect (ASD) is the second most common congenital heart defect with a prevalence of 56/100,000 live births. Higher estimates have been reported more recently, perhaps related to wide availability of echocardiography.[6] ASD is responsible for 15%–20% of all congenital heart defects. They are usually diagnosed incidentally in childhood due to a murmur. Symptoms may develop as age advances. Atrial arrhythmias tend to occur in those over 40 years of age. Spontaneous closure of the defect is rare if defect is >8 mm at birth and after 2–3 years of age. Very rarely, an ASD can enlarge on follow-up.[7,8] 14% of patients with large ASD develop the serious complication of pulmonary vascular disease, usually between 20 and 40 years of age.[9] However, in a series from India, pulmonary vascular disease developed in the first decade in 7% of patients with secundum ASD.[10]

*Types of atrial septal defect*
i. Ostium secundum (~75%)
ii. Ostium primum (15%–20%)
iii. Sinus venosus (5%–10%)
iv. Coronary sinus (<1%)

Patent foramen ovale
Echocardiographic detection of a small defect in fossa ovalis region with a flap with no evidence of right heart volume overload (dilatation of right atrium and right ventricle [RV]). Patent foramen ovale is a normal finding in newborns.

*Diagnostic workup*
i. Clinical assessment
ii. X-ray chest: Right atrial enlargement, right ventricular enlargement, dilated main pulmonary artery segment, and increased pulmonary vasculature markings
iii. Electrocardiogram (ECG): Signs of right ventricular volume overload, seen as incomplete right bundle branch block and right atrial enlargement. Crochetage sign (notched R wave in all inferior limb leads) may be seen in those with large left-to-right shunts.
iv. Echocardiography: This is the key tool not only for the diagnosis of ASD, but also for determining its location and number, for the assessment of pulmonary artery pressure and pulmonary venous drainage, and for evaluating for suitability for device closure of the defect. Transesophageal echo may be required if transthoracic windows are suboptimal.

Transesophageal echo is often performed in the cardiac catheterization laboratory during device closure of ASD.
v. Cardiac catheterization: It is mostly performed for device closure of the defect. A diagnostic catheterization is required in those with pulmonary hypertension and suspected pulmonary vascular disease.

*Indication for closure*
An ASD with left to right shunt, associated with evidence of right ventricular volume overload without evidence of irreversible pulmonary vascular disease (Class I).

Indications for ASD closure remain the same, irrespective of the method of closure.

*Contraindication for closure*
Severe pulmonary arterial hypertension or irreversible pulmonary vascular occlusive disease (Class III). Patients with borderline operability due to pulmonary vascular disease should be referred to a higher center for further evaluation.

**Ideal age of closure**

i. In an asymptomatic child: 2–4 years (Class I). For sinus venosus defect, surgery may be delayed to 4-5 years (Class IIa).
ii. Symptomatic ASD in infancy (congestive heart failure and pulmonary artery hypertension): seen in about 8%–10% of cases. Early closure is recommended (Class I) after ruling out associated lesions (e.g., total anomalous pulmonary venous drainage, left ventricular inflow obstruction, and aortopulmonary window [APW]).
iii. If presenting beyond ideal age: Elective closure irrespective of age as long as there is left-to-right shunt with right heart volume overload and pulmonary vascular resistance (PVR) is within operable range (Class I).
iv. Symptomatic sequel of paradoxical emboli such as stroke or recurrent transient ischemic attack due to transient right-to-left shunting at the atrial level (Class IIa). This complication is more likely with patent foramen ovale with or without atrial septal aneurysm.

**Method of closure**

Surgical: Established mode (Class I).

Device closure: More recent method, used for secundum ASDs only (Class I).

**Device closure is not advised for secundum atrial septal defect (due to higher likelihood of complications) if:**

i. Deficient rims (<5 mm)
ii. Weight <15 kg
iii. Defect size >20 mm/m$^2$.

*Prognosis*

If ASD is closed in childhood, the prognosis is excellent with normal life expectancy. Untreated patients with hemodynamically significant ASD have reduced life expectancy.

*Recommendations for follow-up*

i.  Follow-up after surgery: Clinical and echo in the 1st year only. No further follow-up is required if no residual defect, no pulmonary hypertension, or arrhythmia. Patient/guardians should be explained about reporting to hospital in case of any cardiac symptoms or symptoms suggestive of arrhythmias.

ii.  Follow-up after device closure:

   a.  Antiplatelet agents: Aspirin (3–5 mg/kg/day) is given a day before or immediately after the procedure and then continued;

   •  Device ≤30 mm – Aspirin (3–5 mg/kg/day) is continued for total duration of 6 months.

   •  Device >30 mm – Aspirin (3–5 mg/kg/day) and clopidogrel (1.5–2 mg/kg/day) are given for 3 months followed by aspirin alone for 3 more months.

   b.  Echocardiography: At discharge, 1 month, 6 months, 1 year, and then every 3–5 years.

iii.  Infective endocarditis (IE) prophylaxis is recommended for 6 months after device or surgical closure. However, all patients are advised to maintain good oro-dental hygiene after this period also.

## ISOLATED VENTRICULAR SEPTAL DEFECT

*Background*

Ventricular septal defect (VSD) is the most common congenital heart defect (excluding bicuspid aortic valve [BAV]) and constitutes 20%–30% of all congenital heart defects.[6] The prevalence varies from 3 to 5/1000 live births.[11,12] However, a much higher prevalence (50/1000 live births) is reported due to ease of detection of small muscular VSDs by echocardiography.

Clinical manifestations depend on the size of the defect and the pulmonary and systemic vascular resistances. About 10% of patients with large VSDs die in 1st year, primarily due to congestive heart failure.[13] Rate of spontaneous closure depends on the size and location of the defect. Spontaneous closure is uncommon in large VSDs. Inlet and malaligned VSDs almost never close spontaneously. Muscular VSDs are more likely to close spontaneously, especially if they are not large. Decrease in size of VSD is seen in 25% of patients. Small VSDs have a >50% chance of spontaneous closure by 5 years of age and a >80% chance by adolescence.[14] Progressive right ventricular outflow tract obstruction (Gasul phenomenon) may develop in 13% and aortic regurgitation (AR) in 6% of patients.[15] In the historic series of Dr. Paul Wood, 52% of patients with large VSD developed irreversible pulmonary vascular disease with the onset in infancy in four-fifths of them.[16] The incidence of IE in patients with small VSD is 1.3 per 1000 patient-years.[17]

### Classification of ventricular septal defect

i.  Perimembranous: 80%

ii.  Outlet or subpulmonary (doubly committed): 5%–7%

iii.  Inlet: 5%–8%

iv.  Muscular: 5%–20%, these could be central (midmuscular), apical, marginal (anterior, septal-free wall area), or multiple, "Swiss cheese" type.

### Classification according to the size of the defect

i.  Small (restrictive) VSD: Diameter of the defect is less than one-third of the size of aortic orifice. Right ventricular and pulmonary artery pressure is normal, left-to-right shunt is <1.5:1 and left sided cardiac chambers are normal in size.

ii.  Moderate VSD (restrictive): Diameter of the defect is more than one-third but less than the size of aortic orifice. Right ventricular and pulmonary artery pressure varies from normal to two-thirds of systemic pressure, left-to-right shunt is >1.5:1 and left sided cardiac chambers are dilated.

iii.  Large VSD (nonrestrictive): Diameter of the defect is equal to or more than the size of aortic orifice. Right ventricular and pulmonary artery systolic pressures are systemic or near systemic. Degree of left-to-right shunt depends on PVR. The left-sided cardiac chambers are dilated when PVR is normal or mildly elevated.

### Diagnostic workup

i.  Clinical assessment

ii.  X-ray chest: The X-ray chest may be completely normal if the VSD is small. Cardiomegaly, increased pulmonary vascularity with prominent pulmonary artery segment is suggestive of significant left-to-right flow. The degree of cardiomegaly correlates well with degree of left-to-right shunting. Those with large VSD and significantly elevated PVR show absence of cardiomegaly, prominent pulmonary artery segment, dilated central pulmonary arteries, and decreased pulmonary vascularity in outer third of lung fields.

iii.  ECG: ECG may be normal in small VSD. Patients with significant left-to-right shunt show signs of left ventricular volume overload, left ventricular hypertrophy (tall R waves and tall peaked T waves in inferior and lateral leads with prominent q waves in V5–V6), and left atrial enlargement (broad notched P waves in lead I and II, with a broad deep P terminal force in lead V1). Some cases may show large equidiphasic RS complexes (the Katz-Wachtel pattern) in midprecordial leads. Right ventricular hypertrophy with right-axis deviation, and absence of prominent left ventricular forces, is seen in large

VSD with significantly elevated PVR (Eisenmenger syndrome).

iv. Echocardiography

    a. Transthoracic echo: Defines location, size, number of VSDs, and their relation to atrioventricular (AV) valves and to aortic and pulmonary valves. One can assess pulmonary artery pressure, presence or absence of left heart dilation, aortic cusp prolapse, AR, if present, and associated lesions.

    b. Transesophageal echo: Occasionally required if the transthoracic echo windows are suboptimal. However, transesophageal echo is very useful during device closure of VSD or in the operation room to assess adequacy of closure of VSD.

v. Cardiac catheterization: It is required in patients with pulmonary hypertension and suspected pulmonary vascular disease. Cardiac catheterization is also performed for interventional purpose, in cases undergoing device closure.

## Indications and timing of closure (all Class I recommendations)

I. Small VSD (no symptoms, normal PA pressure, normal left heart chambers, no cusp prolapse):

    a. Annual follow-up till 10 years of age, then every 2–3 years

    b. Closure indicated if the patient has had an episode of endocarditis or develops cusp prolapse with AR or develops progressive significant right ventricular outflow tract obstruction.

II. Moderate VSD:

    a. Asymptomatic (normal pulmonary artery pressure with left heart dilation): Closure of VSD by 2–5 years of age

    b. Symptomatic: If controlled with medications, VSD closure by 1–2 years of age.

III. Large VSD:

    a. Poor growth/congestive heart failure not controlled with medications (furosemide/spironolactone/enalapril ± digoxin): As soon as possible

    b. Controlled heart failure: By 6 months of age.

IV. VSD with aortic cusp prolapse:

    Any VSD with cusp prolapse and directly related AR that is more than trivial: Surgery whenever AR is detected.

All patients with VSD must be advised to maintain good oro-dental hygiene.

### Contraindications for closure

Severe pulmonary arterial hypertension with irreversible pulmonary vascular occlusive disease (Class III).

Patients with borderline operability due to pulmonary vascular disease should be referred to a higher center for further evaluation. The decision to operate or not should be made on an individual basis taking into account the total picture of the case including results of the investigations.

## Method of closure

### Surgery

i. Patch closure is the standard therapy in most patients. Route of closure depends on the location of the defect, but left ventriculotomy is best avoided.

ii. Temporary pulmonary artery banding: Palliative option in patients with-

    a. Multiple VSDs (Swiss cheese VSDs), inaccessible VSDs (Class I)

    b. Patients with contraindications for cardiopulmonary bypass, e.g., sepsis (Class IIa).

iii. Surgical options for patients with borderline operability: Fenestrated VSD patch closure, fenestrated flap valve VSD patch closure, or leaving (or creating) a 5 mm ASD. Such procedures should only be done after discussion with the family as in some cases, pulmonary hypertension may not regress or may regress and later worsen following surgery (Class IIb).

### Device closure

i. Eligibility criteria:

    a. Weight >8 kg (5 kg for muscular VSD)

    b. Left-to-right shunt >1.5:1.

ii. Indications

    a. Class I – Midmuscular VSD, anterior muscular VSD, postoperative residual VSD

    b. Class IIb – Perimembranous VSD with at least 4 mm distance from the aortic valve.

iii. Contraindications

    a. VSD with irreversible pulmonary vascular disease

    b. Preexisting left bundle branch block or conduction abnormalities

    c. Any AR

    d. Associated lesions requiring surgery

    e. Inlet, subpulmonic VSD.

iv. Device should not be deployed if any of the following findings develop at the time of procedure:

    a. Any degree of AR

    b. Conduction defect: complete heart block (CHB)/left bundle branch block

    c. Mitral or tricuspid regurgitation.

## Hybrid procedure (Class IIb)

It can be considered in infants with muscular VSDs where percutaneous approach is prohibitive due to sheath size.

## Recommendations for follow-up

i. Follow-up after surgery: Clinical, ECG, and echo in the 1st year only. No further follow-up is required if no residual defect or pulmonary hypertension.

Patient/guardians should be explained about reporting to hospital in case of any cardiac symptoms or symptoms suggestive of arrhythmias.

ii. Follow-up protocol for device closure:
   a. Antiplatelet agents: Aspirin (3–5 mg/kg/day) is given a day before or immediately after procedure and continued for total duration of 6 months.
   b. Follow-up visits: At 1 month, 6 months, 1 year, then annually till 5 years, and then every 3–5 years. Echocardiogram and ECG are to be done at each visit in addition to clinical evaluation.
   c. IE prophylaxis is recommended for 6 months after device or surgical closure. However, all patients are advised to maintain good oro-dental hygiene after this period also.

## ATRIOVENTRICULAR SEPTAL DEFECT

*Background*

AV septal defects (AVSDs) account for 4%–5% of all congenital heart defects with estimated incidence of 0.19/1000 live births.[6] These lesions can be divided into partial and complete forms. Clinical manifestations and outcome of patients with AVSD depend on the size of VSD, degree of ventricular hypoplasia (if any), AV valve regurgitation, presence or absence of left ventricular outflow tract obstruction, and presence or absence of associated syndromes. Down syndrome is present in 50% of patients with AVSD. Conversely, about 40%–45% of children with Down syndrome have CHD, and AVSD accounts for almost half of these, mostly in its complete form.[18] Patients with Down syndrome tend to develop early and more severe form of pulmonary vascular disease with irreversible changes appearing as early as 6 months of age.[19] The presence of preoperative left AV valve regurgitation is associated with increased risk of surgery and need for reoperations on follow-up. Complete form of AVSD, if left untreated, has a survival of only 54% at 6 months and 35% at 12 months.[20] Partial form of AVSD has a better survival with 50% alive at 20 years of age.[21]

*Types of atrioventricular septal defects*

i. Complete AVSD: Large septal defect with an atrial component (ostium primum defect) and a ventricular component (inlet septal defect), common AV valve ring, and common AV valve. There may be incompetence of the right- and left-sided parts of the common AV valve. Complete form of AVSD is generally associated with large left-to-right shunt, pulmonary artery hypertension, and congestive heart failure.

ii. Partial AVSD: These patients have separate annuli of right and left AV valve. There is a primum ASD. Cleft of the anterior leaflet of AV valve is common with variable degrees of regurgitation.

iii. Intermediate AVSD: Two separate AV valves with primum ASD and small restrictive inlet VSD.

iv. Unbalanced AVSD: One of the ventricular chambers is hypoplastic. This form is usually associated with complex congenital defects such as heterotaxy syndrome (isomerism).

*Diagnostic workup*

i. Clinical assessment

ii. X-ray chest: Cardiomegaly may be present due to dilation of the right or left heart chambers depending on the severity and direction of AV valve regurgitation and the severity and level of left-to-right shunting. Large left-to-right shunts lead to increased pulmonary vascular markings and prominent pulmonary artery conus.

iii. ECG: PR interval prolongation is present in 50% of cases; occasionally, complete AV block develops. Other findings include moderate-to-extreme left-axis deviation, q waves in leads I and aVL (counterclockwise depolarization), and left atrial and ventricular hypertrophy if significant AV valve regurgitation is present. Right ventricular hypertrophy suggests the presence of pulmonary artery hypertension or right ventricular outflow tract obstruction.

iv. Echocardiography: It is the key tool for the diagnosis and assessment of size of atrial and ventricular septal defects, size of the ventricles (balanced or unbalanced), estimation of the pulmonary artery pressures, presence and severity of AV valve regurgitation and for associated lesions such as left superior vena cava, left or right ventricular outflow tract obstruction, and heterotaxy syndrome. Transesophageal echocardiography may be rarely required in older patients with suboptimal transthoracic windows.

v. Cardiac catheterization: Indicated for the assessment of operability in patients with pulmonary hypertension and suspected pulmonary vascular disease.

*Ideal age of surgery*

i. Complete AVSD
   a. Uncontrolled heart failure: Complete surgical repair as soon as possible (Class I)
   b. Controlled heart failure: Complete surgical repair by 3 months of age (Class I)
   c. Pulmonary artery banding: May be considered in select patients under 3 months of age (Class IIb).

ii. Partial or intermediate AVSD, stable, and with normal pulmonary artery pressures: Surgical repair at 2–3 years of age (Class I)

iii. Associated moderate or severe AV valve regurgitation may necessitate early surgery in partial or intermediate forms.

iv. Pulmonary artery banding is reserved for complex cases and in patients with contraindications for cardiopulmonary bypass (Class IIb).

v. Surgery for moderate-to-severe left AV valve regurgitation is recommended as per the guidelines for mitral regurgitation, discussed later (Class I).

vi. Surgery for left ventricular outflow tract obstruction is reasonable with a peak systolic gradient of ≥50 mmHg, or at a lesser gradient if heart failure symptoms are present, or if concomitant moderate-to-severe atrioventricular or aortic regurgitation is present (Class IIa).

vii. Those presenting beyond 6 months of life with significant pulmonary hypertension and suspected elevated PVR should be referred to a higher center for further evaluation to assess operability.

All patients with AVSD must be advised to maintain good oro-dental hygiene.

*Contraindication for surgical repair*
AVSD with severe pulmonary arterial hypertension and irreversible pulmonary vascular occlusive disease (Class III).

Patients with borderline operability due to pulmonary vascular disease should be referred to a higher center for further evaluation. The decision to operate or not should be made on an individual basis taking into account the total picture of the case including results of the investigations.

*Important determinants of long-term prognosis*
These include left AV valve stenosis/regurgitation (5%–10%), subaortic stenosis (5%), atrial arrhythmias, late-onset CHB, and issues related to Down syndrome (if present).

*Recommendations for follow-up*
i. Lifelong follow-up is required.
ii. In patients with no significant residual abnormality, annual follow-up is required till 10 years of age followed by 2–3-yearly follow-up. The patient should undergo physical examination, ECG, and echocardiography at each visit, and a Holter monitor test may be required in select cases.
iii. IE prophylaxis is recommended for 6 months after surgical closure. However, all patients are advised to maintain good oro-dental hygiene after this period also.

## PATENT DUCTUS ARTERIOSUS

*Background*
Patent ductus arteriosus (PDA) constitutes 5%–10% of all congenital heart defects with a prevalence of "symptomatic" PDA being 0.5/1000 live births.[6] Clinical manifestations depend on the diameter and length of PDA and the relative systemic and pulmonary vascular resistances. Frequency of patent ductus increases with decreasing gestational age and decreasing birth weight. Spontaneous closure rate of PDA in preterm neonates varies from 35% to 75% in the 1st year of life.[22] Small PDAs in full-term neonates may close up to 3 months of age, whereas large PDAs are unlikely to

close spontaneously. Rate of spontaneous closure of PDA after infancy is 0.6% per year.[23] In the historic Dr. Paul Wood's series, 79% of patients with large PDA had onset of Eisenmenger syndrome in infancy.[16]

**Classification according to the size of PDA**

i. Large PDA: Associated with significant left heart volume overload (left ventricular end-diastolic dimension >+2Z score for weight), congestive heart failure, and severe pulmonary arterial hypertension. PDA murmur is unlikely to be loud or continuous.

ii. Moderate PDA: Some degree of left heart overload, mild-to-moderate pulmonary artery hypertension, and no/mild congestive heart failure. Murmur is continuous.

iii. Small PDA: Minimal or no left heart overload. No pulmonary hypertension or congestive heart failure. Murmur may be continuous or only systolic.

iv. Silent PDA: Diagnosed only on echo Doppler. These are hemodynamically insignificant, produce no murmur and there is no pulmonary hypertension.

**Diagnostic workup**

i. Clinical assessment

ii. X-ray chest: Cardiomegaly, increased pulmonary vascularity and prominent ascending aorta in those with a large left-to-right shunt. The absence of cardiomegaly with decreased vascularity in the outer third of lung fields and a prominent pulmonary conus indicate elevated PVR.

iii. ECG: There is presence of signs of left ventricular volume overload, hypertrophy [tall R waves and tall peaked T waves in inferior leads (II, III and aVF) and lateral leads (V5-V6), with prominent q waves in V5-V6] and left atrial enlargement (broad notched P waves in leads I and II, with a broad, deep terminal force in lead V1) in those with a large left-to-right shunt. Right ventricular hypertrophy is seen if the PVR is elevated. In small PDA, ECG is normal.

iv. Echocardiography: This is the key tool for the diagnosis of PDA, assessment of its size and anatomy, left atrial and left ventricular dimensions, estimation of the pulmonary artery pressure, and evaluating the suitability of PDA for device closure.

v. Cardiac catheterization: Rarely required for diagnosing a PDA. Cardiac catheterization is indicated in older children and adults with pulmonary hypertension and suspected pulmonary vascular disease. Besides conventional methods to test operability in catheterization laboratory, balloon occlusion of PDA may help decide whether PDA should be closed or not. Those showing pulmonary artery systolic pressure/aortic systolic pressure ratio <0.5 on balloon occlusion testing may be suitable for closure.[24] The main indication for cardiac catheterization currently is for performing device closure of the PDA.

### Ideal age of closure

i. Large/moderate PDA, with congestive heart failure, pulmonary artery hypertension: Early closure (by 3 months) (Class I)
ii. Moderate PDA, no congestive heart failure: 6 months–1 year (Class I). If failure to thrive present, closure can be accomplished earlier (Class IIa).
iii. Small PDA: At 12–18 months (Class I)
iv. Silent PDA: Closure not recommended (Class III)
v. Those presenting beyond 6 months of life with large PDA, significant pulmonary hypertension and suspected elevated PVR should be referred to a higher center for further evaluation to assess operability.

All patients with PDA must be advised to maintain good oro-dental hygiene.

### Contraindication for closure

PDA associated with severe pulmonary arterial hypertension with irreversible pulmonary vascular occlusive disease; and silent PDA (Class III).

### Method of closure

i. Weight >6 kg – Can be individualized. Device closure (preferred as less invasive), coil occlusion, or surgical ligation (Class I).
ii. Weight <6 kg – Surgical ligation (Class I), device/coils (off-label use; Class IIb)
iii. Surgical ligation is recommended in cases of progressively enlarging or symptomatic ductal aneurysm, endarteritis, and PDA with unusual shape not considered suitable for device (Class I).
iv. Drug therapy with indomethacin/ibuprofen/paracetamol not to be used in term babies (Class III).

### Recommendations for follow-up

i. Follow-up after device closure or surgery: Clinical, ECG, and echo in the 1st year only. No further follow-up is required if no residual or associated defect and no pulmonary hypertension. Patient/guardians should be explained about reporting to a hospital in case of any cardiac symptoms.
ii. IE prophylaxis is recommended for 6 months after device or surgical closure. However, all patients are advised to maintain good oro-dental hygiene after this period also.

### PDA in a preterm baby (gestational age <37 weeks)

i. Intervene if baby is in heart failure (small PDAs may close spontaneously).
ii. Approved drugs – indomethacin/ibuprofen/paracetamol (if no contraindication) (Class I).
iii. Mode of drug administration – intravenous or oral. At least two courses of drug therapy should be tried before considering surgical intervention (Class I).

iv. Surgical ligation if above drugs fail or are contraindicated (Class I).
v. Feasibility of device/coil closure of PDA has been demonstrated, but risk of major adverse events is high (Class IIb).
vi. Prophylactic indomethacin or ibuprofen therapy: Not recommended (Class III).

## AORTOPULMONARY WINDOW

### *Background*

APW is a rare congenital cardiac malformation, comprising only 0.1% of all congenital heart defects.[6] In most patients with this anomaly, the defect is moderate to large. Associated anomalies occur in half of the cases and include interrupted aortic arch (most commonly type A interrupted aortic arch), origin of right pulmonary artery from the aorta, coarctation of aorta (CoA), right or left coronary artery origin from main pulmonary artery, tetralogy of Fallot [TOF], etc.[25] Clinical manifestations depend on the diameter of APW, the relative systemic and pulmonary vascular resistances, and associated lesions. Large APW is associated with very early development of advanced pulmonary vascular disease.

### Types of aortopulmonary window[26]

- Type 1 proximal defect (most common): Defect located just above the sinus of Valsalva, with very little inferior aortopulmonary septum above the semilunar valves.
- Type 2 distal defect: Defect located in the uppermost portion of the ascending aorta, with little superior rim of aortopulmonary septum.
- Type 3 total defect: Large defect that spans from semilunar valves to the pulmonary artery bifurcation with little superior and inferior rims.
- Intermediate type: Central defect with adequate superior and inferior rims.

### Diagnostic workup

i. Clinical assessment
ii. X-ray chest: Cardiomegaly with increased pulmonary vascularity is seen in those with significant left-to-right shunt. There is absence of cardiomegaly with decreased vascularity in outer third of lung fields, prominent pulmonary artery segment and dilated central pulmonary arteries in patients with elevated PVR.
iii. ECG: Signs of biventricular hypertrophy in response to volume overload of the left ventricle and pressure overload of the RV. Predominant right ventricular hypertrophy is seen if the PVR is elevated.
iv. Echocardiography: This is the key tool for the diagnosis of APW, assessment of its size and location, relation to semilunar valves and coronary ostia, identification of associated anomalies, assessment

of size of cardiac chambers, and estimation of pulmonary artery pressure.

v.  Cardiac catheterization: Performed for diagnostic purposes in those with pulmonary hypertension and suspected pulmonary vascular disease. Rarely, catheterization is performed for interventional purpose, in those with a small defect suitable for device closure.

vi.  Computed tomography angiography (CTA): Rarely performed as a part of preoperative workup in older children, where details of anatomy are not clear on echocardiography.

### Ideal age of closure

i.  Uncontrolled heart failure: Surgical repair as soon as possible (Class I)

ii.  Controlled heart failure: Elective surgical repair by 3 months of age (Class I)

iii.  In patients with associated anomalies, single-stage repair of all defects is preferred (Class I).

iv.  Those presenting beyond 6 months of life with severe pulmonary hypertension and suspected elevated PVR should be referred to a higher center for further evaluation to assess operability.

All patients with APW must be advised to maintain good oro-dental hygiene.

### Contraindication for closure

Severe pulmonary arterial hypertension with irreversible pulmonary vascular occlusive disease (Class III).

### Method of closure

i.  Surgical patch repair is the treatment of choice (Class I).

ii.  Transcatheter device closure in selected cases of intermediate-type APW (Class IIa).

### Recommendations for follow-up

i.  Follow-up after surgery: Clinical evaluation, ECG, and echo annually till 5 years. No further follow-up is required if no residual defect or pulmonary hypertension. Patient/guardians should be explained about reporting to hospital in case of any cardiac symptoms.

ii.  Lifelong follow-up is required in cases of APW associated with interrupted aortic arch and other important anomalies and in those who had elevated PVR before surgery.

iii.  Antiplatelet recommendation for device closure: Aspirin (3–5 mg/kg/day) is given a day before or immediately after procedure and continued for total duration of 6 months.

iv.  IE prophylaxis is recommended for 6 months after surgical or device closure. However, all patients are advised to maintain good oro-dental hygiene after this period also.

## COARCTATION OF AORTA

*Background*

Coarctation of Aorta (CoA) constitutes 6%–8% of all congenital heart defects with a prevalence of 0.24/1000 live births.[6] It is more common in males. It constitutes 15%–35% of cardiovascular malformations in children with Turner syndrome.[27,28] BAV is the most common associated congenital anomaly seen in 80% of cases.[29]

Clinical presentation depends on the age at presentation. Neonates and infants often present with congestive heart failure. There may be associated aortic arch hypoplasia. Older children and adults present with upper-limb arterial hypertension. The diagnosis may be incidental in older patients while they are being investigated for hypertension. Untreated patients who have survived infancy have a 25% survival at 46 years and 10% survival at 56 years.[30] The risk of residual hypertension and early atherosclerotic cardiovascular disease is increased with late repair.[31] The prevalence of residual hypertension is only 6% in repairs performed between 1 and 5 years of age in comparison with 30%–50% in patients repaired at an older age.[32]

### Diagnostic workup

i.  Clinical assessment: Weak and delayed femoral pulses in comparison with brachial/radial pulse are the key to the diagnosis. The diagnosis may be missed in neonates due to a PDA. Measuring blood pressure in all four limbs shows upper-limb hypertension and is a useful guide to the gradient across the CoA segment.

ii.  X-ray chest: Can be normal in asymptomatic infants and young children. Symptomatic neonates show pulmonary venous congestion with moderate-to-severe cardiomegaly, mainly due to dilation of the RV and the right and left atria. In children and young adults, heart size is normal or only mildly enlarged. Pulmonary vascular markings are normal unless there is an associated left-to-right shunt. The combination of a dilated left subclavian artery proximal to the coarctation and a dilated aorta distal to the coarctation produce a figure of 3 silhouette on the X-ray. Notching of the ribs is a classic radiologic sign caused by collateral flow through dilated, pulsatile posterior intercostal arteries. Rib notching seldom appears before 5–6 years of age. Typical coarctation distal to the left subclavian artery results in bilateral notching of the posterior aspects of third to eighth ribs.

iii.  ECG: Right-axis deviation with right ventricular hypertrophy is the typical finding in symptomatic neonates and infants. The finding of left ventricular hypertrophy in infancy, especially if associated with strain pattern (ST-segment and T wave depression), strongly suggests associated aortic stenosis (AS) or

associated myocardial disease. ECG in older children and adolescents reflects the effects of long-standing left ventricular pressure overload in the form of left ventricular hypertrophy and left atrial enlargement. Prominent, coved ST-segment depression with deeply inverted T waves indicates coexisting BAV with stenosis. Prominent left precordial q waves suggest left ventricular volume overload, which may be due to significant regurgitation through a BAV.

iv. Echocardiography: It provides accurate assessment of site and severity of obstruction, ventricular hypertrophy, ventricular size, and function. Associated findings such as arch hypoplasia, intracardiac defects (such as VSD and BAV), and presence or absence of PDA can be easily diagnosed by echocardiography. Color-flow Doppler assists in localizing the site of obstruction, particularly in cases where two-dimensional (2D) imaging is difficult. Subcostal views in young children show blunted Doppler flow in descending thoracic aorta.

v. CTA/cardiac magnetic resonance imaging (cMRI) may be required in select cases, especially in adults when anatomy is unclear on echo. These tests are also recommended for follow-up imaging in adults with coarctation, who have had surgical or catheter intervention.

vi. Cardiac catheterization: Required if an intervention is planned. Pressure gradient across the CoA segment may be diminished in cases with left ventricular dysfunction and low cardiac output, in the presence of a large PDA, multiple left-sided obstructive lesions in series, or well-developed collateral circulation that decompresses the aorta proximal to coarctation.

## Indications for intervention

i. Patients with CoA gradient ≥20 mmHg (Class I)

ii. Patients of CoA presenting with left ventricular dysfunction, even though the gradient across is <20 mmHg, where left ventricular dysfunction is considered to be due to tight CoA (Class I).

iii. Patients with gradient <20 mmHg, but having upper limb hypertension, left ventricular hypertrophy, or significant collateral formation (Class IIa).

iv. Patients with hypertension who have >50% narrowing at the site of CoA, relative to aortic diameter at diaphragm on CTA/cMRI/angiography, irrespective of pressure gradient (Class IIa).

v. Intervention is not indicated if Doppler gradient across coarctation segment is <20 mmHg with normal left ventricular function and no upper-limb hypertension (Class III).

## Ideal age for intervention

i. With left ventricular dysfunction/congestive heart failure or severe upper-limb hypertension (for age): Immediate intervention (Class I)

ii. Normal left ventricular function, no congestive heart failure, and mild upper-limb hypertension: Intervention beyond 3–6 months of age (Class I)

iii. No hypertension, no heart failure, normal ventricular function: Intervention at 1–2 years of age (Class I)

iv. In patients with significant CoA associated with a sizable VSD, both defects should be repaired in a single stage (Class I).

## Mode of intervention

i. Neonatal presentation: Surgery (Class I). Aortic arch hypoplasia, if associated, should also be repaired.

ii. Critically ill neonates who are considered high risk for surgery (shock-like syndrome and severe left ventricular dysfunction): Balloon angioplasty to tide over the crisis (Class IIa)

iii. Infants with native coarctation: Surgery (Class I) or balloon angioplasty (Class IIa)

iv. Infants with recoarctation: Balloon angioplasty (Class I)

v. Children <25 kg with native coarctation: Balloon angioplasty (Class I) or surgery (Class IIa)

vi. Children <25 kg with recoarctation: Balloon angioplasty ± stenting (Class I)

vii. Children >25 kg and adults with native coarctation: Catheter-based stenting (Class IIa)

viii. Children >25 kg and adults with recoarctation: Catheter-based stenting (Class I)

ix. Elective endovascular stenting of aorta is contraindicated in children <10 years of age (Class III).

## Indications of using a covered stent (provided the anatomy is suitable)

i. Native coarctation where risk of rupture of aorta is high
   a. BAV with ascending aorta dilation
   b. Nearly atretic isthmus (<3 mm diameter)
   c. Turner syndrome
   d. Age >60 years
   e. Marfan syndrome

ii. Recoarctation with aneurysm or pseudoaneurysm at the site of CoA.

All patients with CoA must be advised to maintain good oro-dental hygiene.

## Important determinants of long-term prognosis

Residual or recurring coarctation, status of aortic valve (if bicuspid), aneurysms of the ascending aorta or aneurysm at the intervention site, premature coronary artery disease, and berry aneurysms of the circle of Willis.

## Follow-up recommendations

i. Lifelong follow-up is required.

ii. Annual follow-up initially; later, every 2–3 years if no residual lesions

iii. Clinical assessment should include measuring upper- and lower-limb blood pressure. Echocardiography to be done at each follow-up to exclude any residual issues and to assess for other abnormalities, such as BAV.

iv. Beyond 5 years of age, echo alone may not be sufficient for evaluation. cMRI or CTA is recommended 3–5 yearly or earlier. cMRI is preferable in postsurgical and postballoon angioplasty patients while CTA is preferred after endovascular stenting.

v. Beta-blockers are the preferred drugs for control of hypertension.

vi. IE prophylaxis is needed for 6 months after surgery and intervention. However, all patients are advised to maintain good oro-dental hygiene after this period also.

## AORTIC STENOSIS

### Background

Aortic stenosis (AS) is most often due to stenosis of the aortic valve (80%–85%), but can also be due to obstruction below the valve (subvalvular, 15%, mostly due to discrete membrane) or above the valve (supravalvular, least common). AS is more common in males. The aortic valve can be unicuspid or bicuspid in patients with valvular AS. Patients with unicuspid aortic valve present commonly in neonatal period with critical stenosis, whereas patients with bicuspid valve present more commonly during childhood. BAV has been identified in 1% of the general population; however, the incidence of valvular AS is 0.2–0.4/1000 live births.[6] BAV occurs in 9% of asymptomatic first-degree relatives of patients with BAV.[33] Severity of AS usually progresses in 89% of children under 2 years, and 61% of children over 2 years show progression.[34] Ascending aorta dilation (aortopathy), as defined by a *Z* score >2, has been seen in 74% of children with BAV and the dilatation tends to worsen over time.[35] The incidence of endocarditis in AS is 2.7/1000 patient-years.[36] Untreated aortic valve stenosis presenting in infancy carries a mortality rate of 23%; mortality decreases after that (1.2% per year for the first two decades of life).[37] The risk of sudden death is 0.4%–0.9% per year. Supravalvular stenosis is often associated with Williams–Beuren syndrome.

### Diagnostic workup

i. Clinical assessment

ii. X-ray chest: Normal-sized heart with poststenotic dilation of ascending aorta is seen when obstruction is at valve level. Cardiomegaly indicates severe obstruction with left ventricular dysfunction. More diffuse dilation of aorta indicates associated aortopathy and does not correlate with severity of AS. The cardiac apex may be left ventricular. Pulmonary venous hypertension is seen in severe cases. Adults with valvular AS may show calcification of the valve.

iii. ECG: ECG may be completely normal even in severe AS. Neonates with severe AS may show right ventricular hypertrophy. The presence of left ventricular hypertrophy with ST-segment depression and T wave inversion in the left precordial leads ("strain" pattern) indicates severe AS. Exercise testing can precipitate ST-T changes in asymptomatic patients with severe AS and normal resting ECG. ECG in supravalvular AS can show features of myocardial ischemia due to associated obstruction of coronary ostia.

iv. Echocardiography: It is the key diagnostic imaging technique for assessing the site and severity of AS (peak-to-peak and mean gradients), morphology of the aortic valve, diameter of aortic annulus, evaluation of left ventricular dimensions, mass and systolic function as well as evaluation of associated lesions such as AR, mitral valve disease, and CoA. Transesophageal echocardiography is useful in patients with suboptimal transthoracic window. It is reasonable to screen first-degree relatives of patients with BAV or unicuspid aortic valve with echocardiography for valve disease and aortopathy.

v. CTA/cMRI may be required in older patients with BAV to assess severity of aortopathy and in select cases of supravalvular AS.

vi. Cardiac catheterization: Performed primarily for therapeutic balloon valvuloplasty for valvular AS

vii. Exercise test: May be performed for asymptomatic patients with borderline gradients and a normal ECG. This test should not be done in symptomatic patients.

### Indications and timing of treatment

*Valvular aortic stenosis*

i. Immediate intervention required for:
   a. Newborns with severe AS who are duct dependent (balloon dilation or surgical valvotomy) (Class I)
   b. Infants or children with left ventricular dysfunction due to severe AS, regardless of the valve gradient (Class I).

ii. Elective balloon dilation for:
   a. Asymptomatic or symptomatic patients with AS having gradient by echo-Doppler of >64 mmHg peak or >40 mmHg mean or peak-to-peak gradient of ≥50 mmHg, measured invasively at cardiac catheterization (Class I).
   b. Patients with symptoms due to AS (angina, exercise intolerance) or ECG showing ST-segment changes at rest or during exercise: balloon dilation should be considered for lower gradients (invasively measured) of ≥40 mmHg (Class I).
   c. Asymptomatic child or adolescent with a peak to peak gradient (invasively measured) of ≥40 mmHg, but without ST–T wave changes, if the patient wants to participate in strenuous competitive sports (Class IIb).

iii. Intervention not indicated in asymptomatic children with normal ECG and AS gradient <64 mmHg peak or <40 mmHg mean, by echo-Doppler (Class III).
iv. Balloon dilation should not be performed for AS in the presence of preexisting AR of more than mild severity (Class III).

**Subvalvular aortic stenosis due to discrete membrane: Surgical intervention indicated in**

i. Patients with a peak instantaneous gradient of ≥50 mmHg (Class I)
ii. Patients with a peak instantaneous gradient of <50 mmHg associated with AR of more than mild severity (Class I)
iii. Patients with a peak instantaneous gradient between 30 and 50 mmHg (Class IIb)
iv. Symptomatic patients with a peak instantaneous gradient <50 mmHg in the following situations:
   a. The presence of left ventricular dysfunction attributable to obstruction (Class I)
   b. When pregnancy is being planned (Class IIa)
   c. When the patient plans to engage in strenuous/competitive sports (Class IIa).
v. Intervention not indicated for asymptomatic patients with gradient of <30 mmHg with no or trivial AR (Class III).

Balloon dilation may be attempted in select cases with thin membranes which are away from the aortic valve (Class IIb).

**Supravalvular aortic stenosis: Surgical intervention indicated in**

i. Symptomatic patients with peak instantaneous gradient ≥64 mmHg and/or mean gradient ≥50 mmHg on echo-Doppler (Class I)
ii. Patients with mean Doppler gradient <50 mmHg, if they have any of the following (Class I):
   a. Symptoms attributable to obstruction (exertional dyspnea, angina, and syncope)
   b. Left ventricular systolic dysfunction attributable to obstruction
   c. Severe left ventricular hypertrophy attributable to obstruction
   d. Evidence of myocardial ischemia due to coronary ostial involvement
iii. Asymptomatic patients with mean Doppler gradient ≥50 mmHg may be considered for surgery when the surgical risk is low (Class IIb).

All patients with AS must be advised to maintain good oro-dental hygiene.

**Important determinants of long-term prognosis**

Residual or recurring stenosis, progressive aortic dilation, complications related to prosthetic valve function, such as stuck valve leaflet, paravalvular leak, patient–prosthesis mismatch, and pannus formation. Dysfunction of RV-to-pulmonary artery conduit in those undergoing Ross operation.

**Recommendations for follow-up**

i. All patients with AS require lifelong follow-up irrespective of the type of intervention.
ii. Clinical assessment, ECG, and echo are required, the interval depending on the severity of stenosis.
iii. Those who have undergone a valve replacement, periodic monitoring of anticoagulation (international normalized ratio [INR] levels) is essential. Follow-up after valve interventions should be done annually.
iv. Echocardiography is the mainstay for follow-up for the assessment of aortic valve and ventricular function and above-listed postoperative issues.
v. Patients who have significant AS and are planned for an intervention, should refrain from any sporting activity. Those with asymptomatic moderate stenosis can exercise with low or moderate intensity. Patients with mild degree of stenosis can participate in all sports.
vi. IE prophylaxis is recommended in patients with a prosthetic valve. However, all patients with AS are advised to maintain good oro-dental hygiene.

**PULMONIC STENOSIS**

*Background*

Pulmonary stenosis (PS) is a common congenital heart defect, occurring either as an isolated lesion or in association with other CHD. The prevalence of isolated PS is 7/10,000 and is found in 8%–10% of all patients with CHD.[6,11] The obstruction is at the valve level in 80% to 90% of patients, and the rest have obstruction below or above the pulmonary valve. Isolated subvalvular PS is uncommon, and it is generally associated with a VSD.

The pulmonary valve is dysplastic in 10%–20% of all valvular PS patients.[38] PS may be associated with Noonan, Holt–Oram, or LEOPARD syndrome. Older patients with valvular PS are often asymptomatic, and the diagnosis is made on incidental detection of murmur on routine examination. Occasionally, however, they present in heart failure due to right ventricular dysfunction secondary to severe PS. Neonates and infants with critical PS may present with cyanosis due to right-to-left shunt across an ASD. PS may remain stable, progress, or rarely improve. Progression is more likely in infants than in older children or adults with mild PS.[39,40] The natural history of patients with valvular PS is excellent with 1-year, 2-year, and 15-year actuarial survival rate of 97%, 96%, and 94%, respectively.[41]

**Diagnostic workup**

i. Clinical assessment: Phasic ejection click, a hallmark feature of valvular PS may be absent in dysplastic pulmonary valve.

ii. X-ray chest: This may be normal in patients with mild-to-moderate PS. Prominent pulmonary artery segment due to poststenotic dilation of main and left pulmonary artery localizes the obstruction to valve level. Cardiomegaly with right atrial and right ventricular enlargement indicates right ventricular dysfunction. The pulmonary vascularity is reduced in those with right-to-left shunt at atrial level and in severe cases with reduced cardiac output. The dilated RV is rounded rather than boot shaped (typical of TOF). PS due to dysplastic valve stenosis may not show poststenotic dilation of pulmonary trunk.

iii. ECG: Patients with moderate or severe PS show right-axis deviation and right ventricular hypertrophy. In neonates with critical PS, ECG may show normal QRS axis and left ventricular dominance, especially if the right ventricular cavity is small. Older patients with severe PS may also show right atrial enlargement. R wave amplitude in lead V1 and R/S ratio in leads V1 and V6 correlate with severity of PS. Superior or left-axis deviation may be found in infants with PS who have congenital rubella syndrome or Noonan syndrome.

iv. Echocardiography: It is the key diagnostic tool for assessing the site and severity of PS, morphology of the pulmonary valve, pulmonary annulus diameter, pulmonary valve competence, additional sub- or supravalvar stenosis, evaluation of right ventricular size and function, associated tricuspid regurgitation, other features such as post-stenotic dilation of the main and branch pulmonary arteries, tricuspid valve morphology and shunting across ASD.

v. Cardiac catheterization and angiography: Performed primarily for therapeutic balloon valvuloplasty. Angiography is the gold standard for detailed imaging in patients with peripheral pulmonic stenosis.

vi. CTA/cMRI: Indicated for diagnosis and planning management of patients with peripheral pulmonic stenosis.

**Indications and timing of treatment**

*Valvular pulmonic stenosis*
i. Immediate intervention required for:
   a. Newborns with severe PS who are duct dependent (Class I)
   b. Infants, children, or adults with right ventricular dysfunction due to severe PS, regardless of the valve gradient (Class I)
ii. Elective balloon dilation for:
   a. Asymptomatic or symptomatic patients with valvular PS having peak instantaneous gradient by echo-Doppler of >64 mmHg (Class I)
   b. Neonates and infants with any degree of PS who have mild hypoxia due to mild hypoplasia of RV, even if right ventricular function is normal (Class IIa)

c. Patients with valvular pulmonic stenosis due to dysplastic valve, who meet the above criteria (Class IIa)

**Mode of intervention**

i. Balloon dilatation (Class I)
ii. Surgical intervention reserved only for (Class I):
   a. Subvalvular or supravalvular PS with indications same as in valvular stenosis
   b. Noonan syndrome (dysplastic valve) with hypoplastic annulus
   c. Failed balloon dilatation

**Peripheral pulmonic stenosis**

i. Percutaneous interventional therapy (balloon dilatation ± stenting) is the treatment of choice for focal branch and/or peripheral pulmonary artery stenosis with >50% diameter narrowing, an elevated RV systolic pressure >50 mmHg (or >50% of systemic pressure), difference in perfusion of both lungs of >20% (on lung perfusion scan), and/or symptoms (Class I).
ii. Surgical intervention for the above indications, when stenosis not anatomically amenable to percutaneous interventional therapy (Class I).

**Recommendations for follow-up**

i. All patients with PS require lifelong follow-up.
ii. Clinical assessment, ECG, and echo are required at each visit, the interval depending on the severity of stenosis.
iii. Infants with mild PS in whom intervention is not indicated should be followed 3 monthly until 1 year of age. Thereafter, they should be followed every 1–2 years till 10 years of age and later every 3–5 years. Those with more than mild stenosis (native or after balloon dilation) may be followed every year beyond the infancy period.
iv. IE prophylaxis is recommended in patients with a prosthetic valve. However, all patients with PS are advised to maintain good oro-dental hygiene.

**TETRALOGY OF FALLOT**

*Background*
TOF is the most common cyanotic congenital heart disease with a prevalence of 0.4/1000 live births constituting about 5% of all congenital heart defects.[11] The clinical signs and symptoms seen in infants generally vary in accordance with the degree of right ventricular outflow tract obstruction. Almost two-thirds of newborns with TOF are acyanotic at birth, but by 6 months of age, over half of them have cyanosis at rest.[42] This occurs because of worsening infundibular stenosis which increases right-to-left shunting across the VSD. Intermittent hypercyanotic spells are one of the defining features of TOF. Peak incidence of these episodes occurs

between the 2nd and 6th months of life, and these become infrequent after 2 years of age.[43] Patients with untreated TOF have an estimated 1-year, 3-year, and 10-year survival of 66%, 49%, and 24%, respectively.[44]

### Diagnostic workup

i. Clinical assessment: Degree of cyanosis is the most important aspect of clinical evaluation.
ii. Pulse oximetry: Measuring oxygen saturation by pulse oximeter is recommended at each follow-up examination. Anemia may undermine the severity of clinical cyanosis.
iii. ECG: Typical ECG shows right-axis deviation, right ventricular hypertrophy, and an early QRS transition with abrupt change from an R wave in lead V1 to an rS pattern in lead V2. The presence and depth of Q waves and the amplitude of R waves in leads V5–6 reflects the magnitude of pulmonary blood flow and left ventricular filling. Surgical repair of TOF often disrupts the electrical conduction pathways and >90% of patients exhibit right bundle branch block after surgical repair.
iv. X-ray chest: Normal heart size with an upturned apex ("boot-shaped" heart due to right ventricular hypertrophy), deficiency of the main pulmonary artery segment ("pulmonary bay" seen as a concavity in the upper left cardiac border), and reduced pulmonary vascularity are the cardinal features. Other findings include dilated ascending aorta and right aortic arch (20%–30%).
v. Echocardiography: It is a vital tool for the diagnosis of TOF, and the following features should be noted
   a. Site and degree of right ventricular outflow tract obstruction
   b. Pulmonary valve and annulus size
   c. Size and confluence of branch pulmonary arteries and any evidence of ostial stenosis. McGoon ratio and Nakata index can be calculated to decide suitability for total repair.
   d. Size and location of malaligned VSD and any additional VSD
   e. Coronary anomalies which may interfere with surgical repair (especially anomalous left anterior descending coronary artery crossing the right ventricular outflow tract)
   f. Aortic arch sidedness and branching pattern, aortic dilation, and aortopathy
   g. Additional anomalies such as ASD, complete AVSD, and persistent left superior vena cava
   h. Transesophageal echocardiography may not be required for defining details of anatomy, but it is of great help at the time of surgical repair to check for adequacy of repair.
vi. Laboratory investigation: Hemoglobin (Hb)/packed cell volume (PCV) must be measured in every child with TOF because they require a higher Hb

and Hb of <14 g/dL is considered low. Anemia can precipitate a cyanotic spell. Fluorescence *in situ* hybridization (FISH) for 22q11 deletion may be done if facilities are available. Approximately 20% of TOF patients are FISH positive, and it is recommended to use irradiated blood during surgery in these patients.
vii. CTA: Important for preoperative anatomical delineation, especially in older children where echocardiography may not define the coronary artery anatomy and aorto-pulmonary collaterals.
viii. Cardiac catheterization and angiography: Preoperative cardiac catheterization and invasive angiography have almost entirely been replaced by imaging modalities such as echocardiography, CTA, and, in some cases, cMRI. However, catheterization and angiography may be indicated in select group of patients with suspicion of multifocal pulmonary artery supply, aortopulmonary collaterals, anomalous origin of one pulmonary artery, and anomalous pulmonary venous drainage or in patients where full preoperative information is not possible by echo/CTA. Angiography is frequently performed in postsurgical repair patients who have a residual VSD or pulmonary artery branch stenosis.
ix. cMRI: It is an important imaging tool for follow-up of operated patients, particularly in adolescents and adults with repaired TOF. cMRI is primarily done to monitor the effects of chronic pulmonary regurgitation on right ventricular volume and function and deciding the timing of pulmonary valve replacement in these patients.

### Medical management (Class I)

i. Maintain Hb >14 g/dL (using oral iron or blood transfusion)
ii. Oral propranolol to be given in highest tolerated doses (usual dose is 1–4 mg/kg/day in 2–3 divided doses)
iii. Prostaglandin infusion for neonates with significant cyanosis.

### Management of cyanotic spell

i. Oxygen administration
ii. Place the child in mother's lap in knee-chest position
iii. Intravenous fluid bolus of normal saline at 10–20 ml/kg
iv. Morphine 0.1–0.2 mg/kg intravenously
v. Intravenous metoprolol 0.1 mg/kg over 5 min (can be repeated every 5 min provided no hypotension or bradycardia) or short-acting esmolol infusion (50–200 µg/kg/min)
vi. Sodium bicarbonate 1–2 mEq/kg given (intravenous) after dilution
vii. Blood transfusion if required
viii. For refractory spells: Phenylephrine infusion at 2–5 µg/kg/min, intravenous

ketamine (0.25–1.0 mg/kg bolus dose), intubation, and mechanical ventilation (general anesthesia)

ix. Severe refractory cyanotic spell is an indication for emergency surgery/intervention.

### Timing of surgery

i. All patients need surgical repair

ii. Stable, minimally cyanosed: Total repair at 6–12 months of age or earlier according to the institutional policy (Class I)

iii. Symptomatic children of <6 months of age with significant cyanosis or history of spells despite therapy: Palliation (by systemic-to-pulmonary artery shunt or stenting of the ductus arteriosus/right ventricular outflow tract or pulmonary valve balloon valvuloplasty) or total repair depending on anatomy and center's experience (Class I)

iv. Lower threshold for earlier surgery if no requirement of transannular patch is anticipated

v. Patients having TOF with absent pulmonary valve who are stable: Medical management till 1 year of age followed by total correction with repair of pulmonary artery branch dilation/aneurysm (Class I)

vi. Patients with anomalous left anterior descending artery from the right coronary artery crossing the right ventricular outflow tract, who are likely to need RV-to-pulmonary artery conduit (Class I):
   a. <10 kg weight with significant cyanosis: Aortopulmonary shunt
   b. >10 kg weight: Total repair using conduit, or double-barreled approach after 2 years of age, when the child weighs >10 kg.

### Important determinants of long-term prognosis

These include pulmonary regurgitation (almost invariably present after repair), residual lesions (VSD, right ventricular outflow tract obstruction, and pulmonary artery branch stenosis), right ventricular outflow tract aneurysms, aortic root dilatation, functional abnormalities (right ventricular volume and pressure overload), ventricular dysfunction, conduction abnormalities, and arrhythmias.

### Recommendations for follow-up

i. All patients with TOF require lifelong follow-up in view of the above-listed postoperative issues.

ii. Asymptomatic patients with no residual lesion but with free pulmonary regurgitation, not requiring intervention, should be followed up 1–2 yearly.

iii. Clinical assessment, ECG, and echocardiogram are to be done at each visit. Holter monitoring is indicated in patients suspected of having arrhythmia.

iv. Cardiac catheterization should be performed if any residual lesion is suspected. It may also be required for a percutaneous intervention such as stenting of pulmonary artery branch for stenosis.

v. cMRI is an important investigation for follow-up of these patients. In asymptomatic patients, baseline study should be performed 10 years after surgery with periodic follow-up, with frequency of repeat imaging determined by anatomic and physiological findings. Right ventricular volumes and function assessment by cMRI are an important indicator for pulmonary valve replacement.

vi. Those who have undergone a prosthetic pulmonary valve replacement require periodic monitoring of anticoagulation (INR levels). However, bioprosthetic valves are more commonly used for pulmonary valve replacement and these patients do not require long-term anticoagulation.

vii. IE prophylaxis is indicated in noncorrected patients, patients after surgical repair for 6 months, and patients with percutaneous or surgical pulmonary valve replacement. However, all patients with TOF are advised to maintain good oro-dental hygiene even after 6 months of surgical repair.

### Indications for pulmonary valve replacement[45]

i. Symptomatic patients with symptoms attributed to severe right ventricular volume overload with moderate or severe pulmonary regurgitation (Class I).

ii. Asymptomatic with any two or more of following (Class IIa):
   a. Mild or moderate right ventricular or left ventricular dysfunction.
   b. Severe right ventricular dilation: right ventricular end-diastolic volume >160 ml/m$^2$, right ventricular end-systolic volume >80 ml/m$^2$, or right ventricular end-diastolic volume ≥2 times left ventricular end diastolic volume.
   c. Right ventricular systolic pressure ≥2/3 of systemic pressure due to right ventricular outflow tract obstruction.
   d. Progressive reduction in objective exercise tolerance.

iii. Sustained tachyarrhythmias (Class IIb)

iv. Residual lesions requiring surgical intervention (Class IIb).

## VENTRICULAR SEPTAL DEFECT WITH PULMONARY ATRESIA

### Background

VSD with pulmonary atresia (VSD-PAt) is the most severe form of TOF. In the Bohemia survival study, a prevalence of 0.07 per 1000 live births was observed for VSD-PAt, accounting for 20% of all forms of TOF.[46] About 20% of cases are associated with a syndrome/genetic defect.[47] This CHD usually presents in the neonatal period with cyanosis due to right-to-left shunt at ventricular level. The degree of cyanosis depends on the magnitude of pulmonary blood flow, which in turn, depends on the size of PDA and/or the number and size of aortopulmonary

collaterals. Closure of the ductus in the early neonatal period can be lethal due to acute severe cyanosis. After the neonatal period, cyanosis gradually increases.[6] Rarely, however, patients may present in congestive heart failure due to multiple aortopulmonary collaterals (MAPCAs) or a large PDA.[48] Untreated patients with VSD-PAt have a very dismal outcome with the 1-year and 10-year survival being 50% and 8%, respectively.[44]

*Anatomical types*

VSD-PAt is a complex disease with varying anatomy, especially related to pulmonary artery branches and sources of pulmonary arterial blood supply. The detailed anatomy must be defined in each case to plan management. For the sake of simplicity, VSD-PAt is classified into four types.

- Type A – Short-segment valvular atresia, pulmonary arteries confluent, and good sized, supplied by a PDA
- Type B – Long-segment pulmonary atresia with absent main pulmonary artery. Branch pulmonary arteries confluent and good sized, supplied by a PDA
- Type C – Long-segment pulmonary atresia with absent main pulmonary artery. Branch pulmonary arteries confluent, but pulmonary blood flow dependent predominantly on MAPCAs
- Type D – Long-segment pulmonary atresia with absent main pulmonary artery. Nonconfluent branch pulmonary arteries with MAPCA-dependent pulmonary blood flow.

**Diagnostic workup**

i. Clinical assessment.
ii. Pulse oximetry: Measuring oxygen saturation by pulse oximeter is recommended. Anemia may undermine the severity of clinical cyanosis.
iii. ECG: The findings are same as in TOF; right-axis deviation with right ventricular hypertrophy. Patients with increased pulmonary blood flow may have biventricular hypertrophy and left atrial enlargement.
iv. X-ray chest: The absence of cardiomegaly and pulmonary oligemia as seen in classical TOF. In patients with predominant MAPCA-dependent pulmonary blood flow, pulmonary vascular markings usually have a heterogeneous reticular appearance. The right-sided aortic arch is more common in patients with VSD-PAt (26%–50%) than in those with TOF (20%–30%).
v. Echocardiography: It is a vital tool for the diagnosis; however, it may not delineate the distal pulmonary arterial tree or the sources of pulmonary arterial supply. Hence, additional imaging in the form of cardiac catheterization, CTA/cMRI, or a combination of these is essential for planning definitive repair.
vi. Laboratory investigation: Hb/PCV must be measured in every child because cyanotic patients require a

higher Hb and Hb of <14 g/dL is considered low. Anemia can precipitate a cyanotic spell. FISH for 22q11 deletion may be done if facilities are available. Approximately 25% of patients with VSD-PAt are FISH positive, and it is recommended to use irradiated blood during surgery in these patients.

vii. Cardiac catheterization: Cardiac catheterization is almost always performed in patients with VSD-PAt before planning definitive repair, especially in patients who have had a prior aortopulmonary shunt surgery. It is a Class I indication for Type C and D and Class IIa indication for Type A and B. It helps to assess patency of shunt, confluence of pulmonary artery branches, pulmonary artery pressure, anatomical distribution and size of MAPCAs, proportion of recruitable lung segments, and single or dual supply of lung segments. It is also useful for assessing operability in patients with large MAPCAs presenting late and suspected of having developed pulmonary vascular disease.

viii. CTA is a vital investigation for planning surgery. Either CTA or cMRI is recommended for all patients planned for a repair (Class I). The number of segments of the lung which are supplied by native pulmonary arteries is very well defined by CTA.

ix. cMRI: It provides the same information as CTA without exposure to radiation, but is more time-consuming and is less widely available. It is of particular benefit in the evaluation of operated patients with conduit to monitor the conduit function, assessing right ventricular function and volumes, and deciding timing for further interventions.

**Medical management (Class I)**

i. Neonates presenting with significant cyanosis due to closing ductus should be started on prostaglandin E1 (PGE1) infusion for preoperative stabilization. Lowest maintenance dose should be used once PDA is open.
ii. Maintain Hb >14 g/dL (using oral iron or blood transfusion).
iii. Management of cyanotic spell is the same as described in TOF.
iv. Propranolol to be given in highest tolerated doses (usual dose is 1–4 mg/kg/day in 2–3 divided doses).

**Indications and timing of intervention**

Management depends on the type of VSD-PAt, the institutional experience, and the clinical presentation. In general, this lesion requires a multistage management. Patients with Type C and D have a more complex anatomy and are best referred to a specialized center for further treatment.

- Type A (short-segment VSD-PAt with PDA)
  i. Presentation with significant cyanosis at < 1 year of age: Aortopulmonary shunt (Class I) or PDA stenting (Class IIa) depending on the institutional preference and feasibility.

ii. After 1st intervention or those presenting at ≥1 year of age: Total correction at about 1 year of age, since a RV-to-pulmonary artery (PA) conduit is not required (Class I).

- Type B (long-segment pulmonary atresia with PDA):
  i. Presentation with significant cyanosis at <1 year of age: Aortopulmonary shunt (Class I) or PDA stenting (Class IIa) depending on the institutional preference and feasibility.
  ii. After 1st intervention or in those presenting at ≥1 year of age (Class I):
     a. Optimal pulmonary blood flow with good-sized PAs – Total repair with RV-PA conduit at 3–4 years
     b. Suboptimal pulmonary blood flow with small PAs – additional shunt followed by total repair with RV-PA conduit at 3–4 years
     c. Increased pulmonary blood flow with large PAs – total repair with RV-PA conduit by 1 year

- Type C (long-segment pulmonary atresia with confluent branch pulmonary arteries supplied by MAPCAs) (Class I):
  i. Neonatal presentation – aortopulmonary shunt + unifocalization of MAPCAs or RV-PA conduit keeping VSD open
  ii. After 1st intervention or late presentation
     a. Optimal pulmonary blood flow with good-sized PAs – total repair with RV-PA conduit and VSD closure at 3–4 years
     b. Borderline PAs with large MAPCAs
        i. Unifocalization + RV-PA conduit at 6–12 months
        ii. Total repair with RV-PA conduit and VSD closure at 3–4 years.
     c. Increased pulmonary blood flow and large PAs – single-stage repair (unifocalization of MAPCAs + RV-PA conduit + VSD closure) at about 1 year of age with a preferable weight of >10 kg.

- Type D (long-segment pulmonary atresia with nonconfluent branch pulmonary arteries supplied by MAPCAs) (Class IIa):
  i. Neonatal presentation – Aortopulmonary shunt + unifocalization of MAPCAs.
  ii. After 1st intervention or late presentation
     a. Unifocalization + RV-PA conduit at 6–12 months
     b. Total repair with RV-PA conduit and VSD closure at 3–4 years.

## Some important considerations

- MAPCAs >2 mm in size are suitable for unifocalization.
- The decision regarding type of surgery is also influenced by the number of segments of lung that are supplied by native PAs and/or MAPCAs and, therefore, can be recruited.
- PDA stenting should not be done in the presence of branch pulmonary artery stenosis.
- Additional procedures which may be required in select cases include embolization of aortopulmonary collaterals (only if these lung segments have dual supply), stenting of MAPCAs, and balloon dilatation with or without stenting of branch pulmonary arteries.

### Important determinants of long-term prognosis

These include conduit obstruction, degeneration, pulmonary artery branch stenosis, residual VSD, aortic root dilatation, functional abnormalities (right ventricular volume and pressure overload), ventricular dysfunction, conduction abnormalities, and arrhythmias. Patients who have undergone palliative procedures continue to have cyanosis and may develop complications due to right-to-left shunting.

### Recommendations for follow-up

i. All patients with VSD-PAt require lifelong follow-up.
ii. Annual follow-up in those with no residual lesion.
iii. Palliated patients need to be seen more frequently if their oxygen saturation is low and to decide for the next intervention.
iv. Clinical assessment, ECG, and echocardiogram are required; the interval depending on the nature of repair, residual or additional lesions, symptoms, and functional status. Holter monitoring should be done every 2–3 years even if no arrhythmia is suspected.
v. cMRI is an important investigation for follow-up of these patients to determine the timing of conduit revision and pulmonary valve replacement.
vi. Patients on warfarin, who have undergone a mechanical valve replacement, require periodic monitoring of anticoagulation (INR levels).
vii. IE prophylaxis is indicated in noncorrected or palliated patients with cyanosis, patients after surgical repair for 6 months, and patients with conduits and pulmonary valve replacement. All patients are advised to maintain good oro-dental hygiene even after 6 months of surgical repair.

### Indications for pulmonary valve replacement are same as in tetralogy of Fallot[45]

*Indications for conduit replacement[49]*

i. Symptomatic patients with right ventricular pressure by Doppler >80 mmHg and/or moderate-to-severe pulmonary regurgitation (Class I)
ii. Asymptomatic patients with right ventricular pressure by Doppler >80 mmHg with at least one of the following (Class IIa):
   a. Depressed right ventricular function
   b. Progressive right ventricular dilation

c. Progressive tricuspid regurgitation
d. Sustained atrial or ventricular arrhythmias

## TRANSPOSITION OF GREAT ARTERIES

*Background*

Transposition of great arteries (TGA) is the most common cyanotic CHD at birth, accounting for approximately 5% of all CHDs. Its prevalence is 2–3/10,000 live births.[6] Boys are affected two to three times more as compared to girls. This anomaly is reported to be more common in infants born to mothers with diabetes, poor nutrition, or history of alcohol intake.[50,51] In 70% of cases, there is no associated defect apart from ASD, PDA, or insignificant VSD; these cases are labeled as having simple TGA. Association of TGA with other defects such as large VSD and left ventricular outflow tract obstruction or coarctation is referred to as complex TGA.[52] It is a serious disease and most patients with TGA present very early in life, within few days or weeks after birth. The average life expectancy for an untreated newborn is 0.65 years, with mortality rate at 1 year being close to 90%.[53] With the advent of improved surgical techniques and postoperative care, long-term survival is > 90% with very low reintervention rates.[54] The best surgical option for patients with TGA is an arterial switch operation (ASO) where both arteries are switched so that ventriculoarterial concordance is achieved. However, ASO should be performed early, preferably within 3–4 weeks of life, before the left ventricular mass and volume regresses secondary to fall of PVR after birth. Regression of the left ventricle is accelerated in those with a nonrestrictive ASD. On the other hand, left ventricle does not regress in those associated with a significant sized VSD and/or PDA or left ventricular outflow tract obstruction as it faces elevated pressures.

### Diagnostic workup

i. Clinical assessment: In simple TGA, cyanosis is the dominant feature; no heart murmur is audible in most cases. Those with large VSD and/or PDA present with features of heart failure. Weak femoral pulses indicate associated CoA.
ii. Pulse oximetry and blood gases measurement: Oxygen saturation should be measured in the right arm (preductal), because lower-limb saturation may be higher. In critically ill newborns, arterial (or venous) partial pressure of oxygen should be measured to assess degree of hypoxemia, lactic acidosis, and circulatory failure.
iii. ECG: It shows right-axis deviation and right ventricular hypertrophy, which, however, may be normal for a newborn. Those with large VSD may show good left ventricular forces in addition.
iv. X-ray chest: It can be completely normal or may have narrow pedicle with cardiomegaly (egg-on-side appearance) and increased pulmonary blood flow.

v. Echocardiography: Key diagnostic imaging tool. It shows the following features:
   a. Atrioventricular concordance with ventriculoarterial discordance.
   b. Anterior and rightward position of aorta compared to pulmonary artery.
   c. Site and adequacy of intermixing: size of ASD, VSD, and PDA.
   d. Associated malformations: VSD, left ventricular outflow tract obstruction, coarctation/interruption of aortic arch, and mitral valve abnormalities.
   e. Size of aortic and pulmonary valve annulus and coronary artery origin and course, for planning ASO.
   f. Adequacy of left ventricle to support systemic circulation after an ASO.
vi. Cardiac catheterization: Generally done for performing balloon atrial septostomy. Occasionally, it may be required for the assessment of adequacy of left ventricle for an ASO or to assess PVR in those presenting late.
vii. CTA and cMRI: Rarely performed to clarify anatomy of the aortic arch or to evaluate for a surgically relevant coronary anomaly suspected on echocardiography.

### Indications and timing of surgery

Surgery is indicated for all patients with TGA except in those with irreversible pulmonary vascular disease.

*Presurgical stabilization (Class I):*

i. Is guided by the extent of desaturation and its impact on tissue perfusion. In absence of acidosis and other signs of tissue hypoxemia, an oxygen saturation of >70% is desirable.
ii. Start intravenous infusion of PGE1, soon after delivery, if oxygen saturation is lower than 75% and/or lactic acidosis is present. Monitor respiration as PGE1 infusion may result in apnea. Use lowest maintenance dose once PDA is open.
iii. Balloon atrial septostomy: This procedure is most successful in patients younger than 6 weeks, but can be tried in older infants also if the atrial septum is thin.[55] For older infants with a thicker atrial septum, static balloon dilatation is often successful. It can be done in cardiac catheterization laboratory or at the bedside under echocardiographic guidance. Indications:
   a. Low saturations despite PGE1 infusion and ASD is restrictive (Class I).
   b. Those presenting with low saturation and a restrictive ASD beyond 3–4 weeks with a closed PDA where PGE1 is likely to be ineffective (Class IIa).
   c. Patients with restrictive ASD, not fit for immediate surgery (e.g., having sepsis or respiratory infection) (Class IIa).

d. Restrictive ASD in TGA patients with large VSD or PDA: To decrease left atrial pressure and pulmonary venous hypertension (Class IIa).

*Timing and type of surgery*

i. TGA with intact ventricular septum presenting soon after birth: ASO is the best option (Class I)
Timing of surgery: 7 days to 3 weeks. Surgery indicated earlier, if baby is unstable or has associated persistent pulmonary hypertension of the newborn. Exact timing based on institutional preference, but is best done before 4 weeks.

ii. TGA with intact ventricular septum presenting beyond 3–4 weeks of life with regressed left ventricle:
   a. Presenting between 1 and 2 months: ASO; extracorporeal membrane oxygenation (ECMO) support may be required in some cases (Class IIa)
   b. Presenting between 2 and 6 months: ASO with ECMO support or rapid two-stage ASO[1] or an atrial switch (if rapid two-stage or ECMO not feasible) (Class IIa)
   c. Presenting between 6 months and 2 years: Atrial switch operation (Senning or Mustard operation) (Class IIa). Rapid two-stage ASO[1] to be considered in select cases after detailed evaluation (Class IIb).

iii. TGA with a large VSD and/or a large PDA: ASO with VSD and/or PDA closure by 6 weeks of age (Class I). These patients develop early pulmonary vascular disease and may become inoperable by 6 months to 1 year of age.

iv. TGA with VSD and CoA: ASO with VSD closure and arch repair as soon as possible (Class I). It is preferable to repair all lesions in a single stage.

v. TGA with VSD and significant left ventricular outflow obstruction (Class I):
   a. Subvalvular pulmonary obstruction with normal or near-normal pulmonary valve and pulmonary annulus: ASO with resection of subvalvular stenosis
   b. If obstruction involves pulmonary valve or is subpulmonary but not amenable to resection:

      i. Neonates and infants presenting with significant cyanosis: The options depend on patient's age and surgeon's preference
      a. Systemic-to-pulmonary shunt (at any age) followed by Rastelli-type repair or root translocation (at 2–3 years of age or when the child weighs >10 kg)

b. Réparation à l'Etage Ventriculaire (REV) procedure (usually done at 4–6 months)
c. Pulmonary root translocation (usually done at 6–12 months)
d. Nikaidoh procedure (usually done beyond 6–9 months of age).

ii. In older, stable patients, presenting beyond 2–3 years of age: One of the following surgeries: Rastelli-type repair, Nikaidoh procedure, or root translocation surgery.

c. If the VSD is remote and not amenable to one of the biventricular repairs: Multistage palliative cavopulmonary connection (Class IIa).

## Important determinants of long-term prognosis

Long-term prognosis after surgery depends on the type of surgery performed. Early ASO patients fare well on long term. Attention should be paid to specific issues such as residual defects, coronary insufficiency with resultant myocardial ischemia/ventricular dysfunction, supravalvular obstruction of outflow tracts, neoaortic valve regurgitation, neoaortic root dilatation, arrhythmias, residual pulmonary hypertension, and recurring or late pulmonary hypertension. Specific issues after atrial switch operation include atrial arrhythmias, baffle leaks and obstructions, and development of right ventricular dysfunction.

## Recommendations for follow-up

i. All patients need lifelong follow-up. Follow-up intervals depend on age, type of surgery, and residual findings.
ii. In operated patients with no residual defects: Follow-up visits should be at 1, 3, and 6 months after surgery, yearly after that till onset of adult life, and every 2–3 years thereafter.
iii. Follow-up visits should include clinical assessment, ECG, and echocardiography.
iv. Holter monitoring when suspecting arrhythmias and myocardial ischemia. Frequent Holter monitoring may be required following atrial switch operation.
v. CTA or cMRI for coronary evaluation: Should be done at least once at 5–10 years of age. Earlier and more frequent evaluation of coronary arteries may be done in cases who had intramural coronary artery or difficult coronary transfer at the time of surgery.
vi. IE prophylaxis is recommended in patients with cyanosis; for 6 months after definitive surgery; and in cases with conduits or other prosthetic material. However, all patients are advised to maintain good oro-dental hygiene even after 6 months of definitive surgery.

## DOUBLE-OUTLET RIGHT VENTRICLE

*Background*

Double-outlet RV (DORV) is a condition in which

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1 The first stage of rapid two-stage ASO involves retraining of regressed left ventricle by performing pulmonary artery banding along with the addition of a modified aortopulmonary shunt as the first stage. The same can also be achieved in select patients by stent placement in a PDA (Class IIb). It must be noted that ASO with ECMO support and rapid two-stage ASO have higher morbidity and mortality than primary ASO.

both great arteries are connected completely or predominantly to the RV. By definition, at least more than half of both arteries should arise from the RV.[56,57] The VSD is the only outlet for left ventricle. The VSD could be subaortic (60%–65%), subpulmonary (20%–25%), doubly committed (3%–5%), or remote (noncommitted, 5%).[58] DORV may be associated with a number of cardiac anomalies including PS and CoA. The prevalence of DORV is 0.6/10,000 live births, and it constitutes <1% of all congenital heart defects.[6] The clinical presentation is variable and depends on the location of VSD, presence or absence of obstruction to pulmonary blood flow, and associated cardiac anomalies.[11] Clinical presentation of DORV can be divided into three types:

i. TOF-like presentation: When the VSD is subaortic and there is obstruction to pulmonary outflow, the presentation is with progressive cyanosis and is indistinguishable from classic TOF.
ii. VSD-like presentation: DORV with a large VSD in subaortic location presents with features of heart failure in infancy and mild cyanosis may be present.
iii. TGA-like presentation: The VSD is subpulmonary in location and the great arteries are malposed (Taussig–Bing anomaly). There may be associated coarctation in some cases. Such patients present very early in life with cyanosis and heart failure.

Patients with subaortic or subpulmonary VSD can have total biventricular repair. Those with noncommitted or remote VSD have complex anatomy and may not be suitable for biventricular repair.

**Diagnostic workup**

i. Clinical presentation: Variable as discussed above.
ii. ECG: Nonspecific and depends on the type of DORV. Those with a large subaortic VSD may show biventricular hypertrophy and left ventricular overload pattern. Patients with VSD and PS may have extreme right-axis deviation which differentiates them from classical TOF.
iii. X-ray chest: Cardiomegaly and plethoric lung fields in those with large pulmonary blood flow. Normal-sized heart with oligemic lung fields in TOF type of DORV.
iv. Pulse oximetry: For documenting degree of cyanosis.
v. Echocardiography: It is the primary imaging technique to confirm the diagnosis and to decide the type of surgical repair in a given case. The following features can be defined by echocardiography:
    a. Situs, systemic venous, and pulmonary venous drainage (especially in heterotaxy syndrome).
    b. Location, size, number of VSDs, and their relationship to aortic and pulmonary valve.
    c. Relationship of great arteries to the ventricles and to each other.
    d. Location and severity of PS, status of pulmonary annulus, and valve.
    e. Location and severity of subaortic stenosis, status of aortic annulus, and valve.
    f. Size and competence of AV valves.
    g. Size and function of both ventricles.
    h. Associated defects such as CoA.
vi. Cardiac catheterization: Required in late presenters with pulmonary hypertension suspected of having high PVR. It may also be done in other patients to define the anatomy better.
vii. CTA and cMRI: Associated arch hypoplasia or coarctation may necessitate CTA or cMRI. 3D reconstruction of CT images helps assess routability of VSD to aorta.

**Indication and timing of surgery**

Surgery is indicated for all patients with DORV, except in those with irreversible pulmonary vascular disease.

PGE1 infusion should be started in neonates presenting with duct-dependent pulmonary atresia or severe obstruction to pulmonary outflow.

*Timing and type of surgery depends on double-outlet of right ventricle variant (Class I).*

i. DORV with subaortic VSD and PS (TOF-type DORV):
    a. Presenting with significant cyanosis at <3–4 months: Aortopulmonary shunt.
    b. Presenting with significant cyanosis at >3–4 months: Total repair with closure of VSD and infundibular resection.
    c. Stable patients with no or minimal cyanosis: Total repair with closure of VSD and infundibular resection by 6–12 months.
ii. DORV with large subaortic VSD and pulmonary hypertension (VSD-type DORV):
    a. VSD closure by 6 months of age.
    b. Presenting beyond 6 months of age: Assess for operability and close VSD if operable.
iii. DORV with subpulmonary VSD and pulmonary hypertension (TGA-type DORV):
    a. ASO with VSD closure by 6 weeks of age.
    b. If presenting beyond 3 months, should be evaluated for operability. ASO with VSD closure if operable.
    c. If associated with aortic arch abnormality, arch repair should be done in the same sitting.
iv. DORV with subpulmonary VSD and PS:
    a. If pulmonary obstruction is localized, e.g., subvalvular fibrous membrane or ridge: ASO with resection of subvalvular stenosis.
    b. If pulmonary obstruction is tubular or valvular: One of the following complex surgeries required: Rastelli-type repair, REV procedure, Nikaidoh procedure, or root translocation. A systemic-to-pulmonary artery shunt may be required before these procedures in those presenting early with significant cyanosis. Please refer to the section

on "TGA with VSD and left ventricular outflow tract obstruction" for more details.

v. DORV with remote VSD or associated with other complex anatomy: One should strive to perform biventricular repair by intraventricular baffling of left ventricular connection to aorta. Univentricular palliation is done in cases where biventricular repair is not possible.

### Important determinants of long-term prognosis

The determinants of outcome vary with the anatomical variant and type of surgical correction. Important issues in operated patients include residual lesions, pulmonary valve incompetence, and arrhythmias. Those with RV-to-pulmonary artery conduits develop conduit stenosis and degeneration. Patients who have undergone an ASO may have residual pulmonary hypertension, coronary insufficiency, resultant myocardial ischemia/ventricular dysfunction, supravalvular obstruction of both outflow tracts, neoaortic valve regurgitation, neoaortic root dilatation, and arrhythmias.

### Recommendations for follow-up

i. All patients need lifelong follow-up; frequency to be individualized depending on the type of surgery, presence or absence of residual lesions, and functional status.
ii. If no residual defect after surgery: Annual follow-up till adult life, then every 2 years.
iii. Follow-up visits should include clinical assessment, ECG, and echocardiography.
iv. Holter monitoring every 2–3 years after the age of 5 years.
v. In post-ASO patients, coronary evaluation should be done by CTA or cMRI, at least once after the age of 5 years even if patient is asymptomatic.
vi. IE prophylaxis is recommended in patients with cyanosis and in cases with conduits or other prosthetic material in the heart. Prophylaxis is also required for 6 months after definitive surgery. However, all patients with DORV are advised to maintain good oro-dental hygiene even after 6 months of definitive surgery.

### CONGENITALLY CORRECTED TRANSPOSITION OF GREAT ARTERIES

### Background

Congenitally corrected TGA (ccTGA) is a congenital heart defect characterized by atrioventricular and ventriculoarterial discordance. This double discordance results in physiologically corrected circulation, as the great arteries receive appropriate blood. ccTGA occurs due to abnormal looping of the primitive cardiac tube to the left instead of to the right. ccTGA is a rare condition occurring in 0.3/10,000 live births and constitutes 0.5% of all congenital heart defects.[6] Although the physiology

is corrected, anatomy is not, and morphologic RV supports systemic arterial circulation. In over 90% of cases, ccTGA is accompanied by other cardiac lesions: VSDs in 80%, PS (outflow obstruction to morphologic left ventricle) in 30%–50%, abnormalities of the conduction system in 15%–50%, ASD in 12%, and tricuspid valve abnormalities (as detected at autopsy) in 90%.[59] Mismatch between visceral situs and cardiac position is common, with dextrocardia/mesocardia in one-fourth of patients. The age of presentation is variable, and the presentation is determined by the associated anomalies. Patients present with heart failure if a large VSD is present and as TOF physiology if large VSD is associated with significant PS. The associated anomalies affect the natural history of ccTGA. Approximately 10% are born with complete heart block (CHB). In the rest, the risk of developing CHB is 2% per year and about 30% develop CHB by adulthood.[60] The function of the RV deteriorates after the second decade even in those without any associated anomaly.[61]

### Diagnostic workup

i. Clinical assessment: Features of heart failure, cyanosis, bradycardia, and systolic murmur of tricuspid regurgitation.
ii. Pulse oximetry: For measuring arterial saturation in cyanosed patients.
iii. ECG: In cases with situs solitus, the *P* wave axis is normal and q waves are present in the right precordial leads and absent in the left precordial leads. There may be evidence of CHB and rarely preexcitation pattern may be seen.
iv. X-ray chest: Useful for the assessment of visceral situs and cardiac position. Cardiomegaly with pulmonary plethora suggests associated large VSD. Sometimes, an L-posed aorta produces a bulge on the upper left cardiac border.
v. Echocardiography: Main imaging tool for determining the situs and cardiac position, atrioventricular and ventriculoarterial discordance, relationship of great arteries (aorta anterior and to left of pulmonary artery in situs solitus cases), and associated anomalies. Echocardiography is also very useful for the diagnosis of Ebstein's malformation of left-sided morphologic tricuspid valve, quantification of tricuspid regurgitation and for the assessment of right ventricular function. Older patients may require a transesophageal echo for complete evaluation.
vi. Cardiac catheterization: Generally not required for making the diagnosis. It may have to be performed for demonstrating coronary anatomy or for the measurement of pulmonary artery pressure and PVR in those with large left-to-right shunts.
vii. cMRI: In adults, diagnostic imaging alone with echo could be inconclusive; cMRI can provide the required information. It is very accurate for

evaluating ventricular volume, mass, and function, especially for the systemic RV and for the assessment of coronaries. cMRI is often utilized in follow-up of operated patients after a double-switch operation, for evaluating intra-atrial baffles.

viii. Electrophysiological testing: May be required in selected patients, who have arrhythmia/blocks.

ix. Exercise test: May be performed in a select group of patients to detect early right ventricular dysfunction.

## Indications and timing of surgery[62,63]

The indications and timing of surgery in ccTGA depend on the presence and type of associated anomalies.

*General recommendations*

i. Tricuspid valve (systemic AV valve) surgery for severe regurgitation should be considered before systemic ventricular failure (ejection fraction < 45%) sets in (Class IIa).

ii. Anatomic repair (double-switch operation-atrial switch plus arterial switch or Rastelli) may be considered when left ventricle is functioning at systemic pressure and when such surgery is feasible (Class IIa).

*Indications and timing for specific groups of congenitally corrected transposition of great arteries*

i. No associated anomalies: Medical follow-up to look for any development of tricuspid regurgitation or right ventricular dysfunction (Class I). Neonatal double-switch operation may be considered (Class IIb).

ii. Associated with large VSD:
   a. <3 months: Pulmonary artery banding followed later by double-switch operation (atrial plus arterial switch) (Class I).
   b. >6 months: Double switch (atrial plus arterial switch), provided that the patient has not developed irreversible pulmonary vascular disease (Class I).
   c. 3–6 months: Pulmonary artery banding followed by double-switch operation or direct double-switch operation depending on institutional policy (Class IIa).

iii. Associated with large VSD and left ventricular outflow obstruction (PS):
   a. VSD routable: Double switch (atrial switch plus Rastelli) (Class I); univentricular repair pathway if the surgeon is not comfortable doing double-switch operation and saturation is low (Class IIa).
   b. VSD not routable:
      i. Saturation good – Medical follow-up after informed discussion with family (Class IIa).
      ii. Saturation low – Univentricular repair pathway (Class I).

iv. Associated with complex cardiac malformations: Physiological biventricular repair, root transfer (for nonroutable VSD), or univentricular repair (Class IIa).

v. Associated with CHB: Permanent, dual-chamber pacemaker implantation (Class I).

vi. Associated with progressive development of isolated, severe tricuspid regurgitation or right ventricular dysfunction in a child or adolescent: Cardiac catheterization to measure left ventricular pressure (Class IIa).
   a. If left ventricular pressure is ≥80% of right ventricular pressure: Double-switch operation.
   b. If left ventricular pressure is <80% of right ventricular pressure: pulmonary artery banding to prepare left ventricle.
      i. If left ventricular pressure becomes ≥80% of right ventricular pressure: Double-switch operation.
      ii. If left ventricular pressure remains <80% of right ventricular pressure: Tricuspid valve repair.

vii. Associated with severe tricuspid regurgitation in an adult:
   a. Good right ventricular function, prepared left ventricle: Double-switch operation and tricuspid valve repair (Class IIa).
   b. Good right ventricular function, low left ventricular pressure: Tricuspid valve repair/replacement (Class IIb).

viii. Associated with severe right ventricular dysfunction in an adult: Pulmonary artery banding or cardiac resynchronization therapy or cardiac transplant (Class IIa).

## Important determinants of long-term prognosis

These include right ventricular function, severity of left-sided systemic tricuspid regurgitation, arrhythmias, and cyanosis. Specific issues for operated patients include baffle problems, bradyarrhythmias, tricuspid regurgitation, and issues related to the type of operation done, such as atrial switch, Rastelli procedure, and/or ASO.

## Recommendations for follow-up

i. All patients with ccTGA require lifelong follow-up, usually every year.

ii. Clinical assessment, ECG, and echocardiography should be done at each visit.

iii. Additional imaging may be required for better delineation of anatomy and function in adult patients, best done with cMRI.

iv. Holter monitoring, exercise test, and electrophysiological study may be indicated in select patients.

v. IE prophylaxis is recommended for all patients with cyanosis and in cases with conduits or other prosthetic material in the heart. It is also advised for 6 months after a definitive surgery. However, all patients with ccTGA are advised to maintain good oro-dental hygiene.

## UNIVENTRICULAR HEARTS (SINGLE VENTRICLES)

### Background

Univentricular hearts are defined as the presence of one ventricle instead of two, or the second ventricle is rudimentary without an inlet portion. The main ventricle may be of left (more common) or right ventricular morphology or, in rare cases, indeterminate. The classical examples include double-inlet left ventricle, tricuspid atresia, and hypoplastic left heart syndrome. This term also includes a number of congenital heart defects where two-ventricle (or biventricular) repair is not possible. The examples include unbalanced AVSD, double-outlet RV with a nonroutable VSD, straddling of an AV valve, or a very large/multiple VSDs, not amenable to closure. These defects are often grouped together as "functional univentricular" heart, because the management is on the lines of single ventricle.[64] The prevalence is 2.3/10,000 live births, corresponding to 2.8% of all congenital heart defects.[11,65] Heterotaxy syndrome may be associated in some cases. The single ventricle supports both the systemic and pulmonary circulations. The clinical presentation depends on the ratio of pulmonary-to-systemic blood flow. Infants with unrestricted pulmonary blood flow present with predominant heart failure and mild cyanosis and those with obstruction to pulmonary blood flow have dominant cyanosis and no heart failure. Rarely, there could be a balanced circulation.[6]

### Diagnostic workup

i. Clinical assessment: Helps classify the univentricular heart into increased or restricted pulmonary blood flow group.

ii. ECG: It is not specific and different types of abnormalities are described. It has no specific role, but may detect associated prexcitation, blocks, and atrial arrhythmias.

iii. X-ray chest: Cardiomegaly with pulmonary plethora if the pulmonary flow is unrestricted. Normal-sized heart with oligemic lung fields in those with restricted pulmonary blood flow.

iv. Pulse oximetry: For quantifying cyanosis which may give an estimate of pulmonary blood flow restriction.

v. Echocardiography: Key diagnostic tool. Useful to define anatomy, function of the single ventricle, any restriction to pulmonary blood flow, presence of atrioventricular valve atresia/ regurgitation, assess aortic arch, subaortic obstruction, restrictive atrial septal defect, etc.

vi. Cardiac catheterization: Usually not required for diagnosis. Indications include:
   a. For assessing PVR in patients with unrestricted pulmonary blood flow, presenting beyond 6 months of life.
   b. Patients with unrestricted pulmonary blood flow having an arterial saturation of <90%.
   c. Before bidirectional Glenn procedure, if the patient has undergone a pulmonary artery banding, or if any degree of pulmonary hypertension is suspected.
   d. Before total cavopulmonary connection (Fontan) in all patients.
   e. For interventions such as stenting of ductus arteriosus and occlusion of aortopulmonary collaterals.
   f. Operated patients: Failing Fontan, branch pulmonary artery stenting, fenestration closure or fenestration creation, occlusion of venovenous collaterals.

vii. CTA: Required to define associated lesions such as total anomalous pulmonary venous connection (TAPVC), pulmonary vein stenosis, and systemic venous abnormalities. CTA is frequently done before bidirectional Glenn procedure to assess the adequacy of size of branch pulmonary arteries and to look for any evidence of obstruction or narrowing as that will need repair at the time of surgery. Aortopulmonary and venovenous collaterals are also well seen on CTA.

viii. cMRI: It provides the same information as CTA, without exposing the child to radiation. However, cMRI facility is less commonly available and this test takes longer, sometimes necessitating the use of general anesthesia. It is useful in older children and adults who have undergone Fontan operation as MRI is the gold standard for assessing ventricular volumes and function in a univentricular heart.

ix. Other tests: In those having heterotaxy syndrome, an abdominal ultrasound for situs and polysplenia/asplenia is warranted. A peripheral blood smear for Howell–Jolly bodies should also be done.

### Timing and type of intervention

### Preamble

Surgery for univentricular heart is a palliative procedure. The life expectancy is less than normal (exact age cannot be predicted) and is interposed by interventions over these years.[63] Treating physicians must inform and discuss the details with the parent/guardian before surgery.

The timing and type of intervention depends on age at presentation and presence or absence of obstruction to pulmonary blood flow.

i. Those presenting in neonatal period or within 2–3 months of life (Class I):

a. With increased pulmonary blood flow:
   - Type of surgery: Pulmonary artery banding (usually combined with ligation of PDA).
   - Timing of surgery: At 4–6 weeks of age, preferably before 3 months.
   - Additional procedures may be required if systemic outflow obstruction is present.

b. With decreased pulmonary blood flow (PS group):
- Type of surgery: Systemic-to-pulmonary artery shunt or stenting of ductus arteriosus (depends on institutional policy and clinical scenario).
- Indications of surgery:
  - When systemic arterial saturation is consistently below 70%–75%.
  - In case of pulmonary atresia with duct-dependent pulmonary circulation (baby is usually on prostaglandin infusion).

c. With balanced pulmonary circulation: The baby usually maintains saturations above 80% and is not in failure. Such infants should be followed up closely. If saturation falls below 70% and the age is:
- Below 4–5 months: A systemic-to-pulmonary artery shunt (Class I)
- More than 4–5 months: Bidirectional Glenn procedure if branch pulmonary arteries are of good size (Class I).

ii. Those presenting later in life or have undergone first surgery earlier:
a. With pulmonary hypertension and no PS: Most patients who present beyond 3–4 months would become unsuitable for pulmonary artery banding or any definitive repair in the future due to irreversible increase in PVR. A minority of patients may continue to have low resistance and should be offered pulmonary artery banding after full investigations, which includes cardiac catheterization to test for operability.
b. With normal pulmonary pressure and resistance due to PS/previous pulmonary artery banding/previous aortopulmonary shunt:

- Bidirectional Glenn procedure between 6 and 12 months of age (Class I) (can be considered between 4 and 6 months in certain circumstances).
- Additional source of pulmonary blood flow (not completely interrupting the antegrade flow across pulmonary outflow) can be kept in the presence of adequate ventricular function, no AV valve regurgitation, small-sized pulmonary artery branches, and heterotaxy syndrome.
- Total cavopulmonary connection or completion of Fontan procedure (preferably extracardiac): Between 4 and 7 years of age when the child weighs 15–20 kg. If Fontan required earlier (due to increasing cyanosis or due to pulmonary arteriovenous malformations), a lateral tunnel Fontan can be performed at the age of 3–4 years. Fenestration of Fontan circuit is indicated in high-risk cases.
- Additional procedures such as repair of AV valve regurgitation, atrial septectomy, branch pulmonary artery stenosis repair (plasty), and TAPVC repair may have to be done at the time of bidirectional Glenn procedure or total cavopulmonary connection.

## Important determinants of long-term prognosis

The interventions for a univentricular heart are always palliative, and these patients experience a number of morbidities on long term. These include development of ventricular dysfunction, growth failure, worsening cyanosis, arrhythmias, AV valve regurgitation, thrombosis and thromboembolic events, protein-losing enteropathy, plastic bronchitis, hepatic dysfunction, and chronic Fontan failure.

## Recommendations for follow-up

i. All patients with univentricular heart (operated or unoperated) require lifelong follow-up. Frequency should be individualized, but should be at least once a year in stable cases.
ii. Follow-up after bidirectional Glenn procedure: Clinical assessment, oxygen saturation, ECG, echocardiography, cardiac catheterization, and CTA in select cases to decide suitability and timing of total cavopulmonary connection.
iii. Follow-up after total cavopulmonary connection: Clinical assessment, oxygen saturation, ECG, X-ray chest, and echocardiography. In addition, hematological and kidney and liver function tests should be done. cMRI may be performed at least once in adult life and then repeated, if indicated. Periodic imaging of liver by ultrasound, and additional imaging by CT/MRI/biopsy, especially starting 10 years after total cavopulmonary connection.[66]
iv. Drugs after surgery: Aspirin (3–5 mg/kg/day) for all patients. Oral anticoagulants (warfarin) and sildenafil in select group or as per institution policy.[67]
v. The threshold for performing cardiac catheterization during follow-up should be low as a number of complications can be successfully treated if diagnosed in time.
vi. IE prophylaxis is recommended in patients with cyanosis and in cases with conduits or other prosthetic materials in the heart. However, all patients with univentricular heart are advised to maintain good oro-dental hygiene.

## PERSISTENT TRUNCUS ARTERIOSUS

### Background

Persistent truncus arteriosus is characterized by a single arterial vessel that arises from the base of the heart and gives origin to the coronary, pulmonary, and systemic arteries. It accounts for < 1% of all congenital heart defects with a prevalence rate of 0.07/1000 live births.[46] There is associated microdeletion of chromosome 22q11.2 in about 40% of patients.[68] The aortic arch is right sided in about one-third of patients.[69,70] Aortic arch interruption or coarctation of the aorta is found in 15%–20% of patients with truncus arteriosus.[71] Physiologic consequences of truncus

arteriosus depend on the size of the pulmonary arteries and on the PVR, with truncal valve regurgitation/stenosis adding to the hemodynamic burden on the ventricles. Patients usually present in the first few weeks of life due to congestive heart failure and failure to thrive. Untreated patients have a very high mortality, mainly due to congestive heart failure, with a survival rate of 35% at 6 months and 10% at 1 year.[71]

**Classification of truncus arteriosus[72]**

i. Type A1 – Aorta and main pulmonary artery originate from a single large common trunk.
ii. Type A2 – Both pulmonary arteries arise separately and directly from the truncus.
iii. Type A3 – One pulmonary artery arises from the truncus and the other is supplied by the PDA or collaterals from the aorta.
iv. Type A4 – There is an associated obstructive lesion of the aortic arch.

**Diagnostic workup**

i. Clinical assessment: Little or no cyanosis with bounding pulses in early infancy.
ii. Pulse oximetry: For quantifying cyanosis which may give an estimate of PVR.
iii. X-ray chest: It shows cardiomegaly and increased pulmonary vascular markings. A combination of a right aortic arch and increased pulmonary vascularity is strongly suggestive of truncus arteriosus. A dilated truncal root resembles a dilated ascending aorta. Main pulmonary artery segment may arise at a higher level (Type A1) or may be absent (when pulmonary arteries arise directly from truncus).
iv. ECG: There is usually a normal QRS axis or minimal right-axis deviation and combined ventricular hypertrophy and left atrial enlargement. When pulmonary blood flow is reduced due to increase in PVR, there is right-axis deviation and predominant right ventricular hypertrophy.
v. Echocardiography: It is the key tool for the diagnosis and assessment of anatomy, location of VSD, presence and severity of truncal valve regurgitation or stenosis, and for associated lesions such as aortic arch interruption and coronary artery anomalies.
vi. CTA/cMRI: They are useful in select cases when the anatomy is unclear on echocardiography, especially for the evaluation of aortic arch. These tests are recommended for follow-up imaging after surgical intervention.
vii. Cardiac catheterization: Indicated in older patients beyond infancy with suspected pulmonary vascular disease, for the assessment of operability.
viii. Serum calcium levels and genetic testing for microdeletion of chromosome 22q11.2 when clinically indicated.

*Ideal age for surgery*
Surgery indicated in all, unless the patient is inoperable.
i. Uncontrolled heart failure: Surgical repair as soon as possible (Class I).
ii. Controlled heart failure: Surgical repair by 3–6 weeks of age (Class I).
iii. Bilateral pulmonary artery banding reserved for complex cases and patients with contraindications for cardiopulmonary bypass (Class IIb).

**Type of surgery**

Total repair using RV-to-pulmonary artery conduit. Nonconduit options (Barbero-Marcial technique) may be possible in select cases. The prospects of repeat surgeries in future for conduit obstruction should be discussed with parents. Truncal valve repair is done if truncal valve is regurgitant.

**Contraindication for surgery**

Severe pulmonary arterial hypertension with irreversible pulmonary vascular occlusive disease (Class III). Signs of inoperability include age >1 year, resting systemic arterial saturation <85%, and absence of cardiomegaly.

Patients with borderline operability due to pulmonary vascular disease should be referred to a higher center for further evaluation. The decision to operate or not should be made on an individual basis taking into account the history, examination, and results of all the investigations.

**Important determinants of long-term prognosis**

These include residual or progressive pulmonary hypertension, need for conduit replacement, progressive truncal/neoaortic valve regurgitation, aortic root dilatation/aneurysm, and recurrent arch obstruction in Type A4.

**Recommendations for follow-up after surgery**

i. Lifelong follow-up is required in view of above-listed postoperative issues.
ii. Follow-up after surgery with clinical assessment, X-ray chest, ECG, and echocardiography at 1, 6, and 12 months and yearly thereafter in stable cases.
iii. IE prophylaxis is recommended after surgical repair due to the presence of conduit. All patients are advised to maintain good oro-dental hygiene.

**TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION**

*Background*
TAPVC occurs in 0.5–0.9/10,000 live births and accounts for 1% of all patients born with congenital heart defects.[11,73] It has no specific sex predilection. An ASD is necessary for survival. TAPVC frequently occurs as an isolated lesion, but may be associated with other more complex CHDs such as heterotaxy syndrome with asplenia. TAPVC is classified into four

types depending on the site of drainage.[74] Each type can be obstructive or nonobstructive. The four types and their relative frequencies are: Supracardiac (45%–50%), cardiac (15%–20%), infracardiac (26%–28%), and mixed (where the drainage is at two or more sites, 5%–8%).[75] Obstruction to the drainage of pulmonary veins is most common in the infracardiac variety and least common in the cardiac type. If not treated, TAPVC has a very high mortality with 85% dying in the 1st year of life. The median survival was 2 months (range 1 day to 49 years) in the article published by Hazelrig *et al.*[76] However, some of the exceptional survivors present later in life with clinical features suggestive of a large ASD, but have mild desaturation. Pulmonary venous obstruction significantly reduces median survival from 2.5 months in the nonobstructive group to 3 weeks in the obstructive group.[76] Obstructive TAPVC is the only cyanotic CHD where prostaglandin infusion should be avoided.

### Diagnostic workup

i. Clinical assessment: Depends on whether the obstruction is present or not. Neonates with severe obstruction present with cyanosis and respiratory distress soon after birth, usually within 12 h of birth. Patients with no obstruction present with clinical features of large left-to-right shunts with mild cyanosis.
ii. Pulse oximetry: For quantifying cyanosis which may give an estimate of PVR.
iii. ECG: Right-axis deviation with right ventricular hypertrophy.
iv. X-ray chest: Shows cardiomegaly, prominent pulmonary artery segment, and pulmonary plethora. In older infants and children, one may see a typical figure of 8 or "snowman sign" in cases with supracardiac TAPVC. Neonates with obstructive TAPVC show no or minimal cardiomegaly and severe pulmonary venous hypertension (ground glass appearance).
v. Echocardiography: It is the investigation of choice and gives complete information in majority of patients. The right atrium, RV, and pulmonary artery are dilated. The most important finding is the inability to show connection of pulmonary veins to left atrium and right-to-left shunting through an ASD. The exact site of drainage and the presence or absence of obstruction can be defined, although may be difficult in mixed variety with > 1 site of drainage. Assessment of pulmonary artery pressure is possible with echo-Doppler, and associated anomalies can also be identified.
vi. Cardiac catheterization is very rarely performed. It may be required in patients with pulmonary artery hypertension presenting beyond infancy, where operability is in doubt. An arterial saturation of >85% and a PVR (indexed) of <8 WU.m$^2$ may indicate that the patient is operable.
vii. CTA/cMRI: They are reserved for patients where echocardiography is inconclusive, as in cases with mixed type of TAPVC.

### Indications and timing of surgery (all are Class I recommendations)

i. All patients need surgical repair.
ii. Patients with obstructive TAPVC should undergo emergency surgery.
iii. Surgery should be performed as early as possible in nonobstructive TAPVC, even if they are asymptomatic.
iv. Those presenting late should be evaluated for onset of pulmonary vascular disease and operated if the data suggest operable status.

### Important determinants of long-term prognosis

These include residual pulmonary vein stenosis, residual pulmonary hypertension, progressive stenosis of surgically created anastomosis, and late-onset arrhythmias. Pulmonary venous obstruction occurs in 5%–15% of patients after surgical repair.

### Recommendations for follow-up

i. After surgery, patients should be followed up at 1 month, 6 months, and then annually for 5 years if there is no residual defect (including pulmonary vein obstruction).
ii. ECG and echocardiography should be done at each visit.
iii. CTA/cMRI should be done in operated patients suspected of having pulmonary venous obstruction
iv. Because arrhythmias can occur long after TAPVC surgery, parents/patients should be informed to report if any symptom suggestive of arrhythmia develops beyond 5 years of follow-up.
v. IE prophylaxis is indicated in noncorrected patients and in patients after surgical repair for 6 months. However, all patients with TAPVC are advised to maintain good oro-dental hygiene after this period also.

## EBSTEIN'S ANOMALY OF THE TRICUSPID VALVE

*Background*
Ebstein's anomaly of the tricuspid valve is defined as failure of delamination of tricuspid leaflets from the myocardium. Mostly, the septal and the posterior leaflets are involved, and the anterior leaflet remains mobile. This results in the apical displacement of the functional tricuspid annulus and a rotation toward the outflow tract. A part of the RV inflow gets "atrialized." Tricuspid regurgitation of varying degree is invariably present, and the right atrium is enlarged. Some of the patients may be cyanosed due to right-to-left shunting at the atrial level. Ebstein's anomaly is rare, seen in 1 in 20,000 live births, and forms <1% of all congenital heart defects.[6]

Associated lesions include accessory AV conduction pathways (15%–20%), and an interatrial communication is present in 80%–94% of cases.[77] Patients usually present either in neonatal period or during adolescence or adult life. Neonates may present with cyanosis and heart failure and at times with functional pulmonary atresia with duct-dependent pulmonary circulation. Older patients present with murmur, arrhythmias, or cyanosis. The estimated survival is 76% at 10 years and 53% at 15 years.[78] Prognosis is poor in those diagnosed during fetal or neonatal life. A number of classifications have been described for Ebstein's anomaly; the most commonly used classification was described by Carpentier *et al.* in 1988.[79]

**Diagnostic workup**

i. Clinical assessment.
ii. Pulse oximetry: For quantifying cyanosis.
iii. ECG: It is always abnormal. Ebstein's anomaly can sometimes be diagnosed by a typical ECG showing tall (Himalayan) P waves, prolonged PR interval, right bundle branch block, and deep q waves in leads V1–V4. ECG may show evidence of preexcitation due to accessory AV pathway. Some patients may present with an episode of supraventricular tachycardia. Holter monitoring is done when suspecting an arrhythmia.
iv. X-ray chest: The heart size varies from normal to marked cardiomegaly. The pedicle is narrow, and the cardiac borders are sharp. The heart has a box-like configuration, the right atrium is enlarged, and the lung fields may be oligemic.
v. Echocardiography: It is the key diagnostic tool and shows the following features:
    a. Apical displacement of the septal tricuspid leaflet in the four-chamber view. For diagnosing Ebstein's anomaly, the displacement should be > 8 mm/m$^2$ in adults as some degree of displacement occurs in conditions with right ventricular volume overload.
    b. Severity of tricuspid regurgitation and the velocity of jet
    c. Type of Ebstein's anomaly
    d. Size and function of the functional RV
    e. Presence or absence of ASD and other lesions

Echocardiography also helps in assessing the feasibility of valve repair. Transesophageal echo is rarely required.

vi. Cardiac catheterization: It is rarely performed, unless done for evaluating coronary arteries in older patients (>40 years) undergoing surgical repair of Ebstein's anomaly. Pulmonary artery pressure assessment may be required in those planned for bidirectional cavopulmonary anastomosis (Glenn).
vii. cMRI: Provides quantitative measurement of right ventricular size, volume, and function, which are important for planning surgical repair.

viii. Other tests in select patients: Exercise testing, electrophysiological studies.

**Indications and timing for treatment**

*Neonates*
i. Presenting with significant cyanosis: Intravenous prostaglandin infusion (Class I)
ii. Presenting with heart failure: Diuretics (Class I)
iii. Presenting with arrhythmias: Appropriate antiarrhythmic drug (Class I)
iv. Surgery for those not stabilized with medical therapy (Class IIa).

*Older children and adults*
Tricuspid valve repair (Cone repair) is best done at about 2 years of age for stable cases.
i. Surgery is indicated (Class I) in those with:
    a. Symptoms or deteriorating exercise capacity
    b. Cyanosis (oxygen saturation < 90%)
    c. Paradoxical embolism
    d. Progressive cardiomegaly on chest X-ray (CT ratio > 0.65)
    e. Progressive dilation or dysfunction of the RV on echocardiography.
ii. Symptomatic with arrhythmias: Catheter ablation. Surgery, if not amenable to catheter ablation (Class IIa).

*Types of surgery*
Depends on the underlying anatomy and size of the functional ventricle.

i. Tricuspid valve repair; replacement only if repair cannot be achieved.
ii. Tricuspid valve repair with bidirectional cavopulmonary anastomosis (one and a half ventricle repair).
iii. Single ventricle repair (aortopulmonary shunt/Glenn followed by Fontan surgery).

**Important determinants of long-term prognosis**

These include recurrence of tricuspid valve regurgitation with need for reoperation, right ventricular dilatation and dysfunction, supraventricular and ventricular tachyarrhythmias, and need for pacemaker implantation.

**Recommendations for follow-up**

i. Lifelong follow-up is required for all patients with Ebstein's anomaly.
ii. ECG, X-ray chest, and echocardiography should be done at each visit. Holter, exercise testing, and cMRI may be required in select patients.
iii. Asymptomatic patients who are not candidates for surgery can be followed up every 2–3 years.
iv. Posttricuspid valve repair patients should be followed up every 2–3 years if there is no, trivial or mild tricuspid regurgitation and no other residual defect.

v. Those who have undergone tricuspid valve replacement with a prosthetic valve should be closely monitored by INR testing, for optimal anticoagulation.

vi. Those who have undergone a Glenn or Fontan surgery should have annual follow-up.

vii. IE prophylaxis is indicated in patients who have undergone tricuspid valve replacement, have previous history of endocarditis, or have cyanosis. However, all patients with Ebstein's anomaly are advised to maintain good oro-dental hygiene.

## MITRAL AND AORTIC REGURGITATION

### Background

Mitral (MR) and AR occur most commonly secondary to acute or chronic rheumatic heart disease, and they may coexist in some cases. Congenital MR is uncommon; however, congenital AR due to a BAV is not rare. A proportion of patients with VSD, subaortic stenosis, and TOF develop AR in the course of the disease. These valve lesions can also develop secondary to IE. Mild-to-moderate valve regurgitation has a long asymptomatic period; however, the deterioration is fast once symptoms develop. Dyspnea is a late feature in the course of chronic MR or AR.

### Diagnostic workup

i. Clinical assessment: History and examination.

ii. ECG: May show left ventricular volume overload in severe cases. Atrial fibrillation is seen in long-standing MR patients, is rare in isolated AR.

iii. X-ray chest: The heart size is a good guide to the severity of the lesion. Evidence of pulmonary venous and arterial hypertension denotes severe lesions. Aorta may be dilated in AR secondary to BAV and in other cases of nonrheumatic AR.

iv. Echocardiography: Most useful tool, helping in the assessment of-
   a. Etiology of the valve lesion.
   b. Severity of lesion (very useful in multivalvular cases).
   c. Measurement of left atrial and left ventricular dimensions and left ventricular function.
   d. Suitability of valve for surgical repair.

Measurement of left ventricular dimensions by serial echo-Doppler helps in deciding the timing of valve surgery. The role of 3D echo is expanding and may be performed if facilities are available.

v. Exercise test: In select cases where symptoms are out of proportion to severity of valve lesion.

vi. Other tests: Cardiac catheterization is rarely required. CTA or cMRI may be needed in select cases of AR to define ascending aorta and aortic arch.

### Role of drug therapy

i. No drugs required for asymptomatic cases with mild or moderate MR.

ii. Angiotensin-converting enzyme inhibitors are indicated in patients with severe MR and severe AR. These drugs are usually required over short term before surgery, but may be used long term in patients with symptoms or left ventricular dysfunction who are not candidates for valve surgery and in asymptomatic patients with normal left ventricular systolic function to extend the compensated phase before the need for valve surgery.

iii. Diuretics to be used in those with dyspnea due to heart failure.

iv. Sodium nitroprusside infusion is recommended for the treatment of acute MR; invasive BP monitoring is required for these cases.

v. Anticoagulants (oral) if atrial fibrillation is present.

vi. Secondary prophylaxis, preferably with long-acting benzathine penicillin injection, is required for patients who have underlying rheumatic heart disease as the etiology of MR or AR.

### Indications and timing of surgery

#### Mitral regurgitation[80,81]

i. Symptomatic patients with moderate-to-severe MR with left ventricular ejection fraction >30% (Class I).

ii. Symptomatic patients with moderate-to-severe MR with left ventricular ejection fraction <30% (Class IIb).

iii. Asymptomatic patients with severe MR: Surgery indicated if any of the following present (Class IIa):
   a. Left ventricular ejection fraction <60%
   b. Left ventricular end-systolic dimension Z score >3 for mitral valve replacement and >2.5 if likelihood of mitral valve repair is >95%
   c. Pulmonary artery systolic pressure >50 mmHg.

iv. Asymptomatic patients with moderate or severe MR undergoing cardiac surgery for another indication (Class IIa).

#### Aortic regurgitation[80]

i. Symptomatic patients with moderate-to-severe AR (Class I).

ii. Asymptomatic patients with severe AR: Surgery indicated if any of the following present (Class I):
   a. Left ventricular ejection fraction <50%.
   b. Left ventricular end-systolic dimension Z score >4.

iii. Asymptomatic patients with moderate or severe AR undergoing cardiac surgery for another indication (Class I).

All patients with valvular regurgitation must be advised to maintain good oro-dental hygiene.

### Type of valve surgery[82]

i. Valve repairs are preferable to valve replacements (Class I).

ii. Valve replacement in those in whom valve cannot be repaired (Class IIa).

    a. Ross procedure for young patients with nonrheumatic AR (if expertise available).

    b. Bioprosthetic valve for:
- Female patients planning pregnancy in future.
- Compliance with oral anticoagulation is dubious.

c. Prosthetic metallic valve replacements for the rest of patients.

### Anticoagulation after valve surgery[83]

i. Oral anticoagulant drug: Warfarin or other anticoumarin drug

    a. Desired INR:
- After mitral valve replacement: 3.0 (±0.5)
- After aortic valve replacement: 2.5 (±0.5)
- After valve repair, bioprosthetic valve: 2.5 (±0.5)

    b. Patients should be educated about the importance of maintaining INR in therapeutic range, the effect of diet, medicines, etc., on INR in those taking warfarin and the warning signs of overdose of warfarin. It is desirable that patients carry a Warfarin Card on their person for any emergency management. These patients should be advised to avoid contact sports; otherwise, normal activities are allowed. Regular intramuscular immunization can be given while on oral anticoagulant drugs. Dental surgery is safe with therapeutic levels of INR.

    c. Duration of anticoagulation:
- i. Valve repair, bioprosthetic valve: For 3 months after surgery
- ii. Prosthetic metallic valve: Lifelong.

    d. Oral anticoagulants are also indicated for patients who are in atrial fibrillation.

ii. Aspirin: Dose 3–5 mg/kg/day given in addition to anticoagulation (Class I).

    a. Duration: Valve repair, bioprosthetic valve: For 6 months after surgery

    b. Prosthetic metallic valve: Lifelong

New oral anticoagulant drug (dabigatran) and anti-Xa agents (apixaban and rivaroxaban): Should not be used in place of warfarin/other anticoumarin drugs in patients with prosthetic valves (Class III).

### Recommendations for follow-up

i. Patients with valve lesions require lifelong follow-up

ii. Asymptomatic patients with MR or AR: Clinical assessment, ECG, and echocardiography as per the frequency given below:

    a. Mild MR or AR: Clinical assessment every year, echocardiography every 2 year

    b. Moderate MR or AR: Clinical assessment every 6 months, echocardiography every year

    c. Severe MR or AR: Clinical assessment and echocardiography every 6 months.

More frequent follow-up may be done in patients showing progressive left ventricular dilation.

iii. Postsurgical patients with no residual abnormality: Clinical assessment, ECG, and echocardiography:

    a. Bioprosthetic valve: Every 1–2 years

    b. Post valve repair: Every 1–2 years

    c. Prosthetic metallic valve: Every year. In addition, these patients require frequent monitoring of INR and fluoroscopy (for valve motion).

iv. Postsurgical patients with residual valve abnormality: Follow-up as for native valve regurgitation.

v. IE prophylaxis:[83] All patients must be advised to maintain good oro-dental hygiene after valve surgery. Prophylaxis is reasonable before dental procedures that involve manipulation of gingival tissue, manipulation of the periapical region of teeth, or perforation of the oral mucosa in patients with the following (Class IIa):

    a. Prosthetic heart valves

    b. Prosthetic material used for cardiac valve repair, such as annuloplasty rings and chords

    c. Previous IE

## CARDIAC PACEMAKERS IN CHILDREN

*Background*

The scope and clinical indications of implanting pacemaker and other devices have increased with the current generation of devices being small in size, having a longer battery life, advanced programming capabilities, and ability to treat arrhythmias as well as heart failure. The pacing lead can be put either through a transvenous route (subclavian or axillary vein) or directly over the heart (epicardial pacing). Transvenous pacemaker implantation is preferred approach; however, in specific conditions, epicardial pacemaker is implanted.

### Indications of permanent pacemaker implantation[84]

*Class I indications*

i. Symptomatic patients with advanced second/third-degree AV conduction block

ii. Symptomatic patients with sinus node dysfunction and bradycardia with correlation of symptoms

iii. Postoperative advanced second/third-degree AV conduction block that is not expected to resolve or lasting 10 days after cardiac surgery

iv. Asymptomatic congenital third-degree AV block (CHB) with

    a. Wide QRS escape rhythm

    b. Complex ventricular ectopy

    c. Ventricular dysfunction

d. Slow ventricular rate.
- Age <1 year: Rate <55 beats per minute (bpm) without structural or CHD or rate <70 bpm with structural or CHD
- Age >1 year: Rate <50 bpm without structural or CHD or rate <70 bpm with structural or CHD

e. Prolonged QTc interval
f. Long asystolic pause lasting >3 fold the cycle length of underlying rhythm

*Class IIa indications*

i. Patients with CHD and sinus bradycardia for the prevention of recurrent episodes of intra-atrial reentrant tachycardia
ii. Sinus bradycardia with complex CHD with resting HR <40 bpm or pauses in ventricular rate longer than 3 s
iii. CHD and impaired hemodynamics due to sinus bradycardia or loss of AV synchrony
iv. Unexplained syncope in a patient with prior congenital heart surgery complicated by transient CHB with residual fascicular block, after a careful evaluation to exclude other causes of syncope

*Class IIb indications*

i. Asymptomatic transient postoperative CHB that reverts to sinus rhythm within 10 days, with residual bifascicular block
ii. Congenital CHB in asymptomatic children or adolescent with an acceptable rate, a narrow QRS complex, normal QTc, and normal ventricular function
iii. Asymptomatic sinus bradycardia after biventricular repair of CHD with a resting HR <40 bpm or pauses in ventricular rate >3 s

*Class III indications (pacemaker not required)*

i. Transient postoperative AV block with return of normal AV conduction in otherwise asymptomatic patient
ii. Asymptomatic bifascicular block with or without first-degree AV block after surgery for CHD, in the absence of prior transient complete AV block
iii. Asymptomatic type I second-degree AV block
iv. Asymptomatic sinus bradycardia with longest pause <3 s and a minimum HR >40 bpm

**Epicardial pacemaker is used instead of transvenous endocardial pacemaker in the following circumstances[85]**

i. Weight <10–15 kg
ii. Congenital heart defects, repaired or unrepaired, with right-to-left shunt
iii. Venous abnormality or congenital malformations which make a venous lead implantation impossible,

e.g., tricuspid atresia, post glenn/fontan
iv. After surgical correction of complex cardiac malformation where additional open-heart surgery is required
v. Absence of transvenous access to the chamber to be paced
vi. Mechanical valve in tricuspid valve position

**Pacing mode**

- Atrial-based pacing (AAI[R], VDD, and DDD[R]) is superior to ventricular pacing as it maintains AV synchrony. However, in small children, single-chamber pacing is acceptable.
- In patients undergoing epicardial pacemaker implantation, left ventricular (or systemic ventricle) pacing is superior to right ventricular pacing.

**Other important considerations for cardiac pacing in children**

i. In general, permanent pacemakers are required for life and repeated procedures are necessary to change pacemaker generator and/or pacing leads. The available pacemakers usually have a battery life of 5–10 years depending on the usage.
ii. The procedure should be carried out by experienced operator.
iii. Site of implantation and venous access should be conserved as much as possible.
iv. During transvenous pacing, an additional loop of pacing lead should be left in right atrium in children to allow continued pacing with somatic growth of the child.

**Complications of cardiac pacing in children**

A number of complications secondary to permanent pacemaker have been described; however, some are more likely to occur in children. These include:

i. Lead failure: More frequently seen in children, resulting in inappropriate sensing or capture.
ii. Infection of the pacemaker system is a serious complication and almost always necessitates complete system removal.
iii. Vascular complications: Significant vascular access challenges can also be due to previous cardiac surgical procedures.
iv. Decreased ventricular function secondary to chronic right ventricular pacing over long periods.
v. Others: Pneumothorax, endocarditis/pericarditis, hematoma, atrial or ventricular perforation.

**Recommendations for follow-up after implantation of a pacemaker**

i. Patient to be assessed at the time of discharge, 1–2 weeks, 3 months, and then 6 monthly.
ii. Follow-up should include clinical assessment, including the health of the local site of implantation, ECG, device

interrogation for pacing threshold and battery life, and echocardiography (especially in children with CHD).

iii. X-ray chest should be done annually.

iv. Holter in select cases if indicated.

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## Conflicts of interest

There are no conflicts of interest.

## REFERENCES

1. EUROCAT Steering Committee. Congenital Heart Defect in Europe; c2014. Available from: http://www.eurocat-network.eu/content/ EUROCAT-Special-Report-CHD.pdf. [Last accessed on 2019 Mar 05].

2. Hoffman JI. The global burden of congenital heart disease. Cardiovasc J Afr 2013;24:141-5.

3. Bernier PL, Stefanescu A, Samoukovic G, Tchervenkov CI. The challenge of congenital heart disease worldwide: Epidemiologic and demographic facts. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2010;13:26-34.

4. Warnes CA. The adult with congenital heart disease: Born to be bad? J Am Coll Cardiol 2005;46:1-8.

5. Working Group on Management of Congenital Heart Diseases in India. Consensus on timing of intervention for common congenital heart disease. Indian Pediatr 2008;45:117-26.

6. Allen HD, Shaddy RE, Penny DJ, Feltes TF, Cetta F. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. 9th ed. Philadelphia: Wolters Kluwer; 2016.

7. Radzik D, Davignon A, van Doesburg N, Fournier A, Marchand T, Ducharme G. Predictive factors for spontaneous closure of atrial septal defects diagnosed in the first 3 months of life. J Am Coll Cardiol 1993;22:851-3.

8. McMahon CJ, Feltes TF, Fraley JK, Bricker JT, Grifka RG, Tortoriello TA, *et al.* Natural history of growth of secundum atrial septal defects and implications for transcatheter closure. Heart 2002;87:256-9.

9. Craig RJ, Selzer A. Natural history and prognosis of atrial septal defect. Circulation 1968;37:805-15.

10. Cherian G, Uthaman CB, Durairaj M, Sukumar IP, Krishnaswami S, Jairaj PS, *et al.* Pulmonary hypertension in isolated secundum atrial septal defect: High frequency in young patients. Am Heart J 1983;105:952-7.

11. Hoffman JI, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol 2002;39:1890-900.

12. Tikanoja T. Effect of technical development on the apparent incidence of congenital heart disease. Pediatr Cardiol 1995;16:100-1.

13. Keith JD, Rose V, Collins G, Kidd BS. Ventricular septal defect. Incidence, morbidity, and mortality in various age groups. Br Heart J 1971;33Suppl:81-7.

14. Alpert BS, Mellits ED, Rowe RD. Spontaneous closure of small ventricular septal defects. Probability rates in the first five years of life. Am J Dis Child 1973;125:194-6.

15. Corone P, Doyon F, Gaudeau S, Guérin F, Vernant P, Ducam H, *et al.* Natural history of ventricular septal defect. A study involving 790 cases. Circulation 1977;55:908-15.

16. Wood P. The eisenmenger syndrome or pulmonary hypertension with reversed central shunt. I. Br Med J 1958;2:701-9.

17. Kidd L, Driscoll DJ, Gersony WM, Hayes CJ, Keane JF, O'Fallon WM, *et al.* Second natural history study of congenital heart defects. Results of treatment of patients with ventricular septal defects. Circulation 1993;87:I38-51.

18. Freeman SB, Taft LF, Dooley KJ, Allran K, Sherman SL, Hassold TJ, *et al.* Population-based study of congenital heart defects in down syndrome. Am J Med Genet 1998;80:213-7.

19. Frescura C, Thiene G, Franceschini E, Talenti E, Mazzucco A. Pulmonary vascular disease in infants with complete atrioventricular septal defect. Int J Cardiol 1987;15:91-103.

20. Berger TJ, Blackstone EH, Kirklin JW, Bargeron LM Jr., Hazelrig JB, Turner ME Jr. Survival and probability of cure without and with operation in complete atrioventricular canal. Ann Thorac Surg 1979;27:104-11.

21. Somerville J. Ostium primum defect: Factors causing deterioration in the natural history. Br Heart J 1965;27:413-9.

22. Koch J, Hensley G, Roy L, Brown S, Ramaciotti C, Rosenfeld CR. Prevalence of spontaneous closure of the ductus arteriosus in neonates at a birth weight of 1000 grams or less. Pediatrics 2006;117:1113-21.

23. Campbell M. Natural history of persistent ductus arteriosus. Br Heart J 1968;30:4-13.

24. Zhang DZ, Zhu XY, Lv B, Cui CS, Han XM, Sheng XT, *et al.* Trial occlusion to assess the risk of persistent pulmonary arterial hypertension after closure of a large patent ductus arteriosus in adolescents and adults with elevated pulmonary artery pressure. Circ Cardiovasc Interv 2014;7:473-81.

25. Kutsche LM, Van Mierop LH. Anatomy and pathogenesis of aorticopulmonary septal defect. Am J Cardiol 1987;59:443-7.

26. Jacobs JP, Quintessenza JA, Gaynor JW, Burke RP, Mavroudis C. Congenital heart surgery nomenclature and database project: Aortopulmonary window. Ann Thorac Surg 2000;69:S44-9.

27. Sybert VP. Cardiovascular malformations and

complications in turner syndrome. Pediatrics 1998;101:E11.

28. Prandstraller D, Mazzanti L, Picchio FM, Magnani C, Bergamaschi R, Perri A, *et al.* Turner's syndrome: Cardiologic profile according to the different chromosomal patterns and long-term clinical follow-up of 136 nonpreselected patients. Pediatr Cardiol 1999;20:108-12.

29. Smith DE, Matthews MB. Aortic valvular stenosis with coarctation of the aorta, with special reference to the development of aortic stenosis upon congenital bicuspid valves. Br Heart J 1955;17:198-206.

30. Campbell M. Natural history of coarctation of the aorta. Br Heart J 1970;32:633-40.

31. Maron BJ, Humphries JO, Rowe RD, Mellits ED. Prognosis of surgically corrected coarctation of the aorta. A 20-year postoperative appraisal. Circulation 1973;47:119-26.

32. Liberthson RR, Pennington DG, Jacobs ML, Daggett WM. Coarctation of the aorta: Review of 234 patients and clarification of management problems. Am J Cardiol 1979;43:835-40.

33. Huntington K, Hunter AG, Chan KL. A prospective study to assess the frequency of familial clustering of congenital bicuspid aortic valve. J Am Coll Cardiol 1997;30:1809-12.

34. Kiraly P, Kapusta L, van Lier H, Hofman AO, Daniels O. Natural history of congenital aortic valvar stenosis: An echo and Doppler cardiographic study. Cardiol Young 1997;7:188-93.

35. Warren AE, Boyd ML, O'Connell C, Dodds L. Dilatation of the ascending aorta in paediatric patients with bicuspid aortic valve: Frequency, rate of progression and risk factors. Heart 2006;92:1496-500.

36. Gersony WM, Hayes CJ, Driscoll DJ, Keane JF, Kidd L, O'Fallon WM, *et al.* Bacterial endocarditis in patients with aortic stenosis, pulmonary stenosis, or ventricular septal defect. Circulation 1993;87:I121-6.

37. Campbell M. The natural history of congenital aortic stenosis. Br Heart J 1968;30:514-26.

38. Koretzky ED, Moller JH, Korns ME, Schwartz CJ, Edwards JE. Congenital pulmonary stenosis resulting from dysplasia of valve. Circulation 1969;40:43-53.

39. Lueker RD, Vogel JH, Blount SG Jr. Regression of valvular pulmonary stenosis. Br Heart J 1970;32:779-82.

40. Nugent EW, Freedom RM, Nora JJ, Ellison RC, Rowe RD, Nadas AS. Clinical course in pulmonary stenosis. Circulation 1977;56:I38-47.

41. Samánek M. Children with congenital heart disease: Probability of natural survival. Pediatr Cardiol 1992;13:152-8.

42. Bonchek LI, Starr A, Sunderland CO, Menashe VD. Natural history of tetralogy of fallot in infancy. Clinical classification and therapeutic implications. Circulation 1973;48:392-7.

43. Perloff JK, Marelli AJ. Clinical Recognition of Congenital Heart Disease. 6th ed. Philadelphia: Elsevier Health Sciences; 2012.

44. Bertranou EG, Blackstone EH, Hazelrig JB, Turner ME, Kirklin JW. Life expectancy without surgery in tetralogy of fallot. Am J Cardiol 1978;42:458-66.

45. Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, *et al.* 2018 AHA/ACC guideline for the management of adults with congenital heart disease: Executive summary: A report of the American College of Cardiology/American Heart Association task force on clinical practice guidelines. Circulation 2019;139:e637-97.

46. Samánek M, Vorísková M. Congenital heart disease among 815,569 children born between 1980 and 1990 and their 15-year survival: A prospective Bohemia survival study. Pediatr Cardiol 1999;20:411-7.

47. Ferencz C, Loffredo CA, Correa-Villasenor A, Wilson PD, editors. Malformations of the cardiac outflow tract. In: Genetic and Environmental Risk Factors of Major Cardiovascular Malformations. The Baltimore- Washington Infant Study 1981-1989. Armonk, NY: Futura Publishing; 1997.

48. Danilowicz D, Ross J Jr. Pulmonary atresia with cyanosis. Report of two cases with ventricular septal defect and increased pulmonary blood flow. Br Heart J 1971;33:138-41.

49. Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, *et al.* ESC guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010;31:2915-57.

50. Wren C, Birrell G, Hawthorne G. Cardiovascular malformations in infants of diabetic mothers. Heart 2003;89:1217-20.

51. Yang J, Qiu H, Qu P, Zhang R, Zeng L, Yan H. Prenatal alcohol exposure and congenital heart defects: A meta-analysis. PLoS One 2015;10:e0130681.

52. Sarris GE, Balmer C, Bonou P, Comas JV, da Cruz E, Di Chiara L, *et al.* Clinical guidelines for the management of patients with transposition of the great arteries with intact ventricular septum: The Task Force on Transposition of the Great Arteries of the European Association for Cardio-Thoracic Surgery (EACTS) and the Association for European Paediatric and Congenital Cardiology (AEPC). Cardiol Young 2017;27:530-69.

53. Liebman J, Cullum L, Belloc NB. Natural history of transpositon of the great arteries. Anatomy and birth and death characteristics. Circulation 1969;40:237-62.

54. Khairy P, Clair M, Fernandes SM, Blume ED, Powell AJ, Newburger JW, *et al.* Cardiovascular outcomes after the arterial switch operation for D-transposition of the great arteries. Circulation 2013;127:331-9.

55. Feltes TF, Bacha E, Beekman RH 3rd, Cheatham JP, Feinstein JA, Gomes AS, *et al.* Indications for cardiac catheterization and intervention in pediatric cardiac disease: A scientific statement from the American Heart Association. Circulation 2011;123:2607-52.

56. Anderson RH, Becker AE, Wilcox BR, Macartney FJ, Wilkinson JL. Surgical anatomy of double-outlet right ventricle – A reappraisal. Am J Cardiol 1983;52:555-9.

57. Anderson RH, McCarthy K, Cook AC. Continuing medical education. Double outlet right ventricle. Cardiol Young

2001;11:329-44.

58. Sridaromont S, Ritter DG, Feldt RH, Davis GD, Edwards JE. Double-outlet right ventricle. Anatomic and angiocardiographic correlations. Mayo Clin Proc 1978;53:555-77.

59. Dyer K, Graham TP. Congenitally corrected transposition of the great arteries: Current treatment options. Curr Treat Options Cardiovasc Med 2003;5:399-407.

60. Huhta JC, Maloney JD, Ritter DG, Ilstrup DM, Feldt RH. Complete atrioventricular block in patients with atrioventricular discordance. Circulation 1983;67:1374-7.

61. Dimas AP, Moodie DS, Sterba R, Gill CC. Long-term function of the morphologic right ventricle in adult patients with corrected transposition of the great arteries. Am Heart J 1989;118:526-30.

62. Tweddell JS. What do we really know about the management of patients with congenitally corrected transposition of the great arteries? J Thorac Cardiovasc Surg 2017;154:1023-5.

63. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, *et al.* ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: A report of the American College of Cardiology/American Heart Association task force on practice guidelines (Writing committee to develop guidelines on the management of adults with congenital heart disease). Developed in collaboration with the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol 2008;52:e143-263.

64. Jacobs ML, Mayer JE Jr. Congenital heart surgery nomenclature and database project: Single ventricle. Ann Thorac Surg 2000;69:S197-204.

65. Khairy P, Poirier N, Mercier LA. Univentricular heart. Circulation 2007;115:800-12.

66. Rychik J. The relentless effects of the fontan paradox. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2016;19:37-43.

67. Alsaied T, Alsidawi S, Allen CC, Faircloth J, Palumbo JS, Veldtman GR. Strategies for thromboprophylaxis in fontan circulation: A meta-analysis. Heart 2015;101:1731-7.

68. Marino B, Digilio MC, Toscano A. Common arterial trunk, DiGeorge syndrome and microdeletion 22q11. Prog Pediatr Cardiol 2002;15:9-17.

69. Butto F, Lucas RV Jr., Edwards JE. Persistent truncus arteriosus: Pathologic anatomy in 54 cases. Pediatr Cardiol 1986;7:95-101.

70. Marcelletti C, McGoon DC, Danielson GK, Wallace RB, Mair DD. Early and late results of surgical repair of Truncus arteriosus. Circulation 1977;55:636-41.

71. Freedom RM, Yoo SJ, Mikailian H, Williams WG. The Natural and Modified History of Congenital Heart Disease. New York: Wiley-Blackwell; 2003.

72. Van Praagh R, Van Praagh S. The anatomy of common aorticopulmonary trunk (truncus arteriosus communis) and its embryologic implications. A study of 57 necropsy cases. Am J Cardiol 1965;16:406-25.

73. Ferencz C, Rubin JD, McCarter RJ, Brenner JI, Neill CA, Perry LW, *et al.* Congenital heart disease: Prevalence at livebirth. The Baltimore-Washington infant study. Am J Epidemiol 1985;121:31-6.

74. Craig JM, Darling RC, Rothney WB. Total pulmonary venous drainage into the right side of the heart; report of 17 autopsied cases not associated with other major cardiovascular anomalies. Lab Invest 1957;6:44-64.

75. Seale AN, Uemura H, Webber SA, Partridge J, Roughton M, Ho SY, *et al.* Total anomalous pulmonary venous connection: Morphology and outcome from an international population-based study. Circulation 2010;122:2718-26.

76. Hazelrig JB, Turner ME, Blackstone EH. Parametric survival analysis combining longitudinal and cross-sectional – Censored and interval-censored data with concomitant information. Biometrics 1982;38:1-15.

77. Danielson GK, Driscoll DJ, Mair DD, Warnes CA, Oliver WC Jr. Operative treatment of Ebstein's anomaly. J Thorac Cardiovasc Surg 1992;104:1195-202.

78. Attie F, Rosas M, Rijlaarsdam M, Buendia A, Zabal C, Kuri J, *et al.* The adult patient with Ebstein anomaly. Outcome in 72 unoperated patients. Medicine (Baltimore) 2000;79:27-36.

79. Carpentier A, Chauvaud S, Macé L, Relland J, Mihaileanu S, Marino JP, *et al.* A new reconstructive operation for Ebstein's anomaly of the tricuspid valve. J Thorac Cardiovasc Surg 1988;96:92-101.

80. Carapetis JR, Beaton A, Cunningham MW, Guilherme L, Karthikeyan G, Mayosi BM, *et al.* Acute rheumatic fever and rheumatic heart disease. Nat Rev Dis Primers 2016;2:15084.

81. Johnson JT, Eckhauser AW, Pinto NM, Weng HY, Minich LL, Tani LY. Indications for intervention in asymptomatic children with chronic mitral regurgitation. Pediatr Cardiol 2015;36:417-22.

82. Finucane K, Wilson N. Priorities in cardiac surgery for rheumatic heart disease. Glob Heart 2013;8:213-20.

83. Nishimura RA, Otto CM, Bonow RO, Carabello BA, Erwin JP 3rd, Fleisher LA, *et al.* 2017 AHA/ACC focused update of the 2014 AHA/ACC guideline for the management of patients with valvular heart disease: A report of the American College of Cardiology/American Heart Association task force on clinical practice guidelines. J Am Coll Cardiol 2017;70:252-89.

84. Sumitomo N. Device therapy in pediatric and congenital heart disease patients. J Arrhythm 2014;30:428-32.

85. Singh HR, Batra AS, Balaji S. Cardiac pacing and defibrillation in children and young adults. Indian Pacing Electrophysiol J 2013;13:4-13.

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