



Original Article

Guidelines for the management of common congenital heart diseases in India: A consensus statement on indications and timing of intervention[☆]



Anita Saxena^{a,*}, Jay Relan^b, Ravi Agarwal^c, Neeraj Awasthy^d, Sushil Azad^e, Manisha Chakrabarty^f, Kulbhushan S. Dagar^d, Velayoudam Devagourou^g, Baiju S. Dharan^h, Saurabh K. Gupta^g, Krishna S. Iyer^e, M. Jayranganathⁱ, Raja Joshi^j, B.R.J. Kannan^k, Ashish Katewa^l, Vikas Kohli^f, Shyam S. Kothari^g, K.M. Krishnamoorthy^h, Snehal Kulkarni^m, Rohit Manoj Kumarⁿ, Raman Krishna Kumar^o, Sunita Maheshwari^p, Krishna Manohar^q, Ashutosh Marwah^r, Smita Mishra^r, Smruti R. Mohanty^m, Kona Samba Murthy^s, Nageswara Rao Koneti^t, P.V. Suresh^u, S. Radhakrishnan^e, Palleti Rajashekar^g, Sivasubramanian Ramakrishnan^g, Nitin Rao^v, Suresh G. Rao^m, Chinnaswamy H.M. Reddy^p, Rajesh Sharma^r, Krishnanaik Shivaprakasha^w, Raghavan Subramanyan^x, R. Suresh Kumar^y, Sachin Talwar^g, Munesh Tomar^z, Sudeep Verma^{aa}, Vijayakumar Raju^{ab}

^a Convener, All India Institute of Medical Sciences, New Delhi, India

^b Writing Committee, All India Institute of Medical Sciences, New Delhi, India

^c Madras Medical Mission, Chennai, India

^d Max Super Speciality Hospital, New Delhi, India

^e Fortis Escorts Heart Institute, New Delhi, India

^f Apollo Hospitals, New Delhi, India

^g All India Institute of Medical Sciences, New Delhi, India

^h Sree Chitra Tirunal Institute for Medical Sciences & Technology, Trivandrum, Kerala, India

ⁱ Jayadeva Institute of Cardiovascular Sciences and Research, Bangalore, Karnataka, India

^j Sir Ganga Ram Hospital, New Delhi, India

^k Vadamalayan Hospitals, Madurai, Tamil Nadu, India

^l Sri Sathya Sai Sanjeevani Hospital, Raipur, Chhattisgarh, India

^m Kokilaben Dhirubhai Ambani Hospital, Mumbai, Maharashtra, India

ⁿ Postgraduate Institute of Medical Education and Research, Chandigarh, India

^o Amrita Institute of Medical Sciences, Kochi, Kerala, India

^p Narayana Institute of Cardiac Sciences, Bangalore, Karnataka, India

^q Sri Sathya Sai Sanjeevani International Centre for Child Heart Care and Research, Palwal, Haryana, India

^r Jaypee Hospital, Noida, Uttar Pradesh, India

^s Innova Children's Heart Hospital, Hyderabad, Telangana, India

^t Care Hospital, Hyderabad, Telangana, India

^u Narayana Hrudayalaya, Bangalore, Karnataka, India

^v Star Hospital, Hyderabad, Telangana, India

^w H.N. Reliance Hospital, Mumbai, Maharashtra, India

^x Frontier Lifeline Hospital, Chennai, India

^y Believers International Heart Centre, Thiruvalla, Kerala, India

^z Nutema Hospital, Meerut, Uttar Pradesh, India

^{aa} Krishna Institute of Medical Sciences, Secunderabad, Telangana, India

^{ab} GKNM Hospital, Coimbatore, Tamil Nadu, India

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* Corresponding author. Department of Cardiology, All India Institute of Medical Sciences, New Delhi, 110029, India. Fax: 011 26588663.

E-mail address: anitasaxena@hotmail.com (A. Saxena).

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ABSTRACT

Introduction: A number of guidelines are available for management of congenital heart diseases 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 congenital heart diseases, as often these patients present late in the course of the disease and may have co-existing morbidities and malnutrition.

Process: Guidelines emerged following expert deliberations at the National Consensus Meeting on Management of Congenital Heart Diseases in India, held on the 10th and 11th of August, 2018 at the All India Institute of Medical Sciences.

Objectives: The aim of the study was to frame evidence-based guidelines for (i) indications and optimal timing of intervention in common congenital heart diseases and (ii) follow-up protocols for patients who have undergone cardiac surgery/catheter interventions for congenital heart diseases.

Recommendations: Evidence-based recommendations are provided for indications and timing of intervention in common congenital heart diseases, including left-to-right shunts, obstructive lesions, and cyanotic congenital heart diseases. In addition, protocols for follow-up of postsurgical patients are also described.

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1. 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 per 1000 live births.^{2,3} If access to screening, early diagnosis, and treatment is available, more than 90% of patients born with CHD survive to adult life with good long-term outcomes.⁴ Most middle- and low-income countries lack such an advanced level of care for children with CHD. Considering birth prevalence as 9 per 1000, the estimated number of children born with CHD every year in India approximates 2,40,000, posing a tremendous challenge for the families, society, and health-care system.

2. Justification for developing Indian guidelines

Evidence-based recommendations for 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 the antenatal period and often present late in the course of the disease. These patients are often underweight and malnourished and have co-morbidities such as recurrent infections and anemia. Many of the late presenters have an advanced level of pulmonary hypertension, ventricular dysfunction, hypoxia, polycythemia, and so on. Modifications in the treatment protocol may be required for optimizing the outcomes in such patients. All these factors justify the need for separate guidelines for management of CHD in India, including the timing of intervention.

A statement on “consensus on timing of intervention for common congenital heart disease” that originated from a Meeting of Working Group on Management of Congenital Heart Disease in India was published in the year 2008.⁵ This statement was revised and updated in a subsequent National Consensus Meeting, which was held in New Delhi after a gap of 10 years, in August 2018. Considering the growing population of postoperative patients, including those needing regular follow-up, we added protocols for follow-up of these patients.

3. Preamble

1. Every pediatrician/cardiologist/other health-care provider must strive to get a complete diagnosis on a patient suspected of having heart disease, with the help of a higher center, if needed.

2. The proposed guidelines are meant to assist health-care providers (pediatrician, cardiologist, pediatric cardiologist) for managing cases with CHDs in their practice. Although 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 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.

4. Aims and objectives

The aim of the study was to outline the optimal timing of intervention in common CHDs and to formulate guidelines and protocols for follow-up of patients who have undergone surgery/catheter interventions for CHD.

5. Guidelines for individual congenital heart defects

5.1. Atrial septal defect

Background

Atrial septal defect (ASD) is the second most common congenital heart defect with a prevalence of 56 per 100,000 live births.⁶ The defect is usually diagnosed incidentally; symptoms may develop as age advances. Atrial arrhythmias tend to occur in those older than 40 years. Spontaneous closure of the defect is rare if the defect is > 8 mm at birth, and after 2–3 years of age.⁷ Fourteen percent of patients with large ASD develop the serious

complication of pulmonary vascular disease, usually between 20 and 40 years of age.⁸ Ostium secundum ASD (75%) is the commonest type. Other types include ostium primum (15–20%), sinus venosus (5–10%), and coronary sinus (<1%) defect.

Diagnostic workup

The workup includes clinical assessment, X-ray of the chest, electrocardiogram (ECG), and echocardiography. Transesophageal echo may be required if transthoracic windows are suboptimal. Diagnostic catheterization is performed in those with pulmonary hypertension and suspected pulmonary vascular disease.

Indication for closure

An ASD with a left-to-right shunt associated with evidence of right ventricular volume overload (class I). Indications for ASD closure remain the same irrespective of the method of closure.

Contraindications for closure

Irreversible pulmonary vascular occlusive disease (class III) is a contraindication for closure. 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, surgery is indicated at 2–4 years of age (class I). For sinus venosus defect, surgery may be delayed to the age of 4–5 years (class IIa).
- II. In case of symptomatic ASD in infancy (congestive heart failure and pulmonary arterial hypertension), early closure is recommended (class I) after ruling out associated lesions (e.g., total anomalous pulmonary venous drainage, left ventricular inflow obstruction, aortopulmonary window [APW], and so on).
- III. If presenting beyond the ideal age, elective closure is indicated irrespective of age as long as there is left-to-right shunt with right heart volume overload and pulmonary vascular resistance (PVR) is within the operable range (class I).

Method of closure

Surgical closure is the established method of closure (class I). Device closure is a more recent method, used for secundum ASDs only (class I).

Device closure is not advised for secundum ASDs if rims are deficient (<5 mm), and in those weighing <15 kg (owing to higher likelihood of complications).

Recommendations for follow-up

- I. With regard to follow-up after surgery, clinical evaluation and echocardiography are to be performed in the first year only. No further follow-up is required if there is no residual defect, pulmonary hypertension, or arrhythmia. The patient/guardian should be explained about reporting to the hospital in case of any cardiac symptoms, or symptoms suggestive of arrhythmias.
- II. Follow-up after device closure is as follows:
 - a. Antiplatelet agents: aspirin (3–5 mg/kg/day) should be given a day before or immediately after the procedure and then continued.
 - i. Device \leq 30 mm: aspirin (3–5 mg/kg/day) should be continued for a total duration of 6 months.
 - ii. Device >30 mm: aspirin (3–5 mg/kg/day) and clopidogrel (1.5–2 mg/kg/day) should be given for 3 months, followed by aspirin alone for 3 more months.
 - b. Echocardiography should be performed at discharge, 1 month, 6 months, 1 year, and then every 3–5 yearly.

- 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.

5.2. Isolated ventricular septal defect

Background

Ventricular septal defect (VSD) is the most common congenital heart defect (excluding the bicuspid aortic valve [BAV]); its prevalence varies from 3 to 5 per 1000 live births.^{9,10} Clinical manifestations depend on the size of the defect and the pulmonary and systemic vascular resistances. Some of the small- and moderate-sized VSDs can close spontaneously. In the historic series of Dr. Paul Wood,¹¹ 52% of patients with large VSD developed irreversible pulmonary vascular disease with the onset in infancy in four-fifths of them. The perimembranous region (80%) is the commonest site of VSD; the outlet or subpulmonary (5–7%), inlet (5–8%), and muscular (5–20%) regions are the other sites.

VSDs are best classified according to the size of the defect:

- I. Small (restrictive) VSD: The diameter of the defect is less than one-third the size of the aortic orifice. Right ventricular and pulmonary artery (PA) pressures are normal, the left-to-right shunt is < 1.5:1, and the size of left-sided cardiac chambers is normal.
- II. Moderate VSD (restrictive): The diameter of defect is more than one-third but less than the size of the aortic orifice, right ventricular and PA systolic pressure varies from normal to two-thirds of systemic pressure, the left-to-right shunt is > 1.5:1, and left-sided cardiac chambers are dilated.
- III. Large VSD (nonrestrictive): The diameter of the defect is equal to or more than the size of the aortic orifice, and right ventricular and PA systolic pressures vary from systemic to near-systemic pressures. The degree of the left-to-right shunt depends on PVR. The left-sided cardiac chambers are dilated when PVR is normal or mildly elevated.

Diagnostic workup

The workup includes clinical assessment, X-ray of the chest, ECG, and echocardiography. Cardiac catheterization may be required in patients with pulmonary hypertension and suspected pulmonary vascular disease or for interventional purpose.

Indications and timing of closure (all class I recommendations)

- I. For patients with small VSD (no symptoms, normal PA pressure, normal left heart chambers, and no cusp prolapse), recommendations are as follows:
 - a. Annual follow-up is needed until 10 years of age and then every 2–3 yearly.
 - b. Closure is indicated if the patient has had an episode of endocarditis, develops cusp prolapse with aortic regurgitation (AR), or develops progressive significant right ventricular outflow tract obstruction.
- II. For patients with moderate VSD, recommendations are as follows:
 - a. When asymptomatic (normal PA pressure with left heart dilation), closure of VSD is indicated by 2–5 years of age.

- b. When symptomatic, but controlled with medications, VSD closure is indicated by 1–2 years of age.
- III. For patients with large VSD, recommendations are as follows
 - a. In case of poor growth/congestive heart failure not controlled with medications, closure is indicated as soon as possible.
 - b. In case of controlled heart failure, closure is indicated by 6 months of age.
- IV. VSD with aortic cusp prolapse
 - In case of any VSD with cusp prolapse and directly related AR that is more than trivial, surgery is indicated whenever AR is detected.

All patients with VSD must be advised to maintain good oral hygiene.

Contraindications for closure

Severe pulmonary arterial hypertension with irreversible pulmonary vascular occlusive disease (class III) is a contraindication for closure. 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.

Modes of VSD closure

Surgery

- I. Patch closure is the standard therapy in most patients.
- II Pulmonary artery banding may be considered as a palliative option for patients with multiple VSDs (Swiss cheese VSDs) and inaccessible VSDs (class I) and for patients with contraindications for cardiopulmonary bypass, for example, sepsis (class IIa).

Device closure

- I. Eligibility criteria are as follows: weight >8 kg (5 kg for muscular VSD) and left-to-right shunt >1.5:1.
- II. Indications are as follows: muscular VSD and postoperative residual VSD (class I) and perimembranous VSD with at least 4-mm distance from the aortic valve (class IIb).
- III. Contraindications for device closure are as follows: Pre-existing left bundle branch block or conduction abnormalities, AR of any degree, associated lesions requiring surgery, and inlet or subpulmonic VSD.
- IV. Device should not be deployed if the patient develops any AR, conduction defect, or mitral or tricuspid regurgitation at the time of procedure.

Recommendations for follow-up

- I. Clinical evaluation, ECG, and echocardiography are to be performed in the first year only. No further follow-up is required if there is no residual defect or pulmonary hypertension. The patient/guardian should be explained about reporting to the hospital in case of any cardiac symptoms or symptoms suggestive of arrhythmias. However, for those who underwent device closure, a long-term follow-up is recommended to look for late-onset complications such as conduction abnormalities, AR, and so on.

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

5.3. Atrioventricular septal defect

Background

Atrioventricular septal defects (AVSDs) account for 4–5% of all congenital heart defects. Clinical manifestations and outcome of patients with AVSD depends on the size of VSD, the degree of ventricular hypoplasia (if any), atrioventricular valve regurgitation, the presence or absence of left ventricular outflow tract obstruction, and the presence or absence of associated syndromes. Down syndrome is present in 50% of patients with AVSD, and these patients tend to develop an early and more severe form of pulmonary vascular disease.¹² The complete form of AVSD, if left untreated, has a survival of only 54% at 6 months and 35% at 12 months.¹³ The partial form of AVSD has a better survival of 50% at 20 years of age.¹⁴

Types of AVSD

- I. Complete AVSD: It is characterized by large septal defect with an atrial component (ostium primum defect) and a ventricular component (inlet septal defect), common atrioventricular valve ring, and common atrioventricular valve. It is generally associated with a large left-to-right shunt, pulmonary arterial hypertension, and congestive heart failure.
- II. Partial AVSD: It is characterized by primum ASD with separate annuli of the right and left atrioventricular valve.
- III. Intermediate AVSD: This condition is characterized by two separate atrioventricular valves with primum ASD and a small restrictive inlet VSD.
- IV. Unbalanced AVSD: One of the ventricular chambers is hypoplastic. It is usually associated with complex congenital defects such as heterotaxy syndrome (isomerism).

Diagnostic workup

The workup includes clinical assessment, X-ray of the chest, ECG, and echocardiography. Cardiac catheterization is indicated for assessment of operability in patients with pulmonary hypertension and suspected pulmonary vascular disease.

Ideal age of surgery

- I. Complete AVSD
 - a. In case of uncontrolled heart failure, complete surgical repair is indicated as soon as possible (class I).
 - b. In case of controlled heart failure, complete surgical repair is indicated by 3 months of age (class I).
 - c. Pulmonary artery banding may be considered in select patients younger than 3 months of age (class IIb).
- II. Partial or intermediate AVSD (stable and with normal PA pressures): Surgical repair is indicated at 2–3 years of age (class I).
- III. Associated moderate or severe atrioventricular valve regurgitation may necessitate early surgery in partial or intermediate forms.
- IV. Those presenting beyond the age of 6 months 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 oral hygiene.

Contraindication for surgical repair

AVSD associated with severe pulmonary arterial hypertension and irreversible pulmonary vascular disease (class III) is a contraindication for surgical repair.

Important determinants of long-term prognosis

These include left atrioventricular valve stenosis/regurgitation (5–10%), subaortic stenosis (5%), atrial arrhythmias, late-onset complete heart block (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 until 10 years of age, followed by 2–3 yearly follow-up. The patient should undergo physical examination, ECG, and echocardiography at each visit; Holter monitoring 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.

5.4. Patent ductus arteriosus

Background

Patent ductus arteriosus (PDA) constitutes 5–10% of all congenital heart defects.⁶ Clinical manifestations depend on the diameter and length of PDA and the relative systemic and pulmonary vascular resistances. Small PDA in full-term neonates may close up to 3 months of age, whereas a large PDA is unlikely to close spontaneously. In the historic work by Dr. Paul Wood,¹¹ 79% of patients with large PDA had onset of Eisenmenger syndrome in infancy.

PDA is best classified according to the defect size: large-sized (significant left heart volume overload and severe pulmonary arterial hypertension with or without congestive heart failure), moderate-sized (some degree of left heart volume overload and mild to moderate pulmonary arterial hypertension), and small-sized (minimal or no left heart overload and no pulmonary hypertension). Very small PDA diagnosed only on echo-Doppler is hemodynamically insignificant, and it is labeled as a silent PDA.

Diagnostic workup

The workup includes clinical assessment, X-ray of the chest, ECG, and echocardiography. Cardiac catheterization is rarely required for operability assessment in older children and adults with pulmonary hypertension and suspected pulmonary vascular disease. It is usually performed as a part of the device closure procedure. Computed tomographic angiography (CTA) is sometimes performed in adults if the suitability of PDA anatomy for device closure is not apparent on echocardiography.

Ideal age of closure

- I. In case of large/moderate PDA with congestive heart failure and pulmonary arterial hypertension, early closure is indicated (by 3 months of age) (class I).
- II. In case of moderate PDA (no congestive heart failure), closure is indicated between the age of 6 months and 1 year (class I). If failure to thrive is present, closure can be accomplished earlier (class IIa).
- III. In case of small PDA, closure is indicated at the age of 12–18 months (class I).

- IV. Those presenting beyond the age of 6 months with large PDA, significant pulmonary hypertension, and suspected elevated PVR, should be referred for operability assessment to a higher center.

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. In those weighing >6 kg, the method of closure can be individualised: device closure (preferred as less invasive), coil occlusion, or surgical ligation (class I).
- II. In those weighing <6 kg, the method of closure can be as follows: surgical ligation (class I) and device/coils (off-label use; class IIb).

Recommendations for follow-up

- I. Follow-up after device closure or surgery: Clinical evaluation, ECG, and echocardiography are to be performed in the first year only. No further follow-up is required if there is no residual defect and no pulmonary hypertension. The patient/guardian 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.

5.5. Aortopulmonary window

Background

APW is a rare anomaly, comprising 0.1% of all congenital heart defects.⁶ It is associated with other anomalies in half of the cases, most commonly type A interrupted aortic arch.¹⁵ 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. APW can be proximal, distal, or a combination of both.¹⁶

Diagnostic workup

The workup includes clinical assessment, X-ray of the chest, ECG, and echocardiography. Cardiac catheterization is performed for diagnostic purposes in those with severe pulmonary hypertension and suspected pulmonary vascular disease. CTA is performed for older children and adults, where anatomical details are not clear on echocardiography.

Ideal age of closure

- I. In case of uncontrolled heart failure, surgical repair should be done as soon as possible (class I).
- II. In case of controlled heart failure, defect should be electively repaired surgically by 3 months of age (class I).
- III. Those presenting beyond the age of 6 months 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 oral hygiene.

Contraindication for closure

Severe pulmonary arterial hypertension with irreversible pulmonary vascular disease (class III) is a contraindication for closure.

Method of closure

Surgical patch repair is the treatment of choice (class I). Transcatheter device closure can be performed in case of small defects with adequate rims all around (class IIa).

Recommendations for follow-up

- I. Follow-up after surgery: Clinical evaluation, ECG, and echocardiography are to be performed annually until 5 years. No further follow-up is required if there is no residual defect or pulmonary hypertension. The patient/guardian should be explained about reporting to the hospital in case of any cardiac symptoms.
- II. Lifelong follow-up is required in those with APW who have residual pulmonary hypertension and in those who had elevated PVR before surgery.
- III. IE prophylaxis is recommended for 6 months after surgical or device closure. However, all patients are advised to maintain good oral hygiene after this period also.

5.6. Coarctation of the aorta

Background

Coarctation of the aorta (CoA) constitutes 6–8% of all congenital heart defects.⁶ It is more common in males. BAV is the commonest associated congenital anomaly, seen in 80% of cases.¹⁷ Clinical presentation depends on the age at presentation. 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.¹⁸ The risk of residual hypertension and early atherosclerotic cardiovascular disease is increased with late repair.¹⁹ The prevalence of residual hypertension is only 6% in patients who undergo repair between 1 and 5 years of age in comparison with 30–50% in patients who undergo repair at an older age.²⁰

Diagnostic workup

The workup includes clinical assessment, X-ray chest, ECG, and echocardiography. CTA/cardiac magnetic resonance imaging (cMRI) may be required in select cases, especially in adults when anatomy is unclear on echocardiography and for follow-up after surgical or catheter intervention. Cardiac catheterization is primarily performed for catheter intervention.

Indications for intervention

The following are the indications for intervention:

- I. Patients with CoA gradient ≥ 20 mmHg (class I).
- II. Patients with CoA presenting with left ventricular dysfunction, even though the CoA gradient 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 the aortic diameter at the level of

diaphragm on CTA/cMRI/angiography, irrespective of pressure gradient (class IIa).

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

Ideal age for intervention

- I. In patients with left ventricular dysfunction/congestive heart failure or severe upper limb hypertension (for age), immediate intervention (class I) is indicated.
- II. In patients with normal left ventricular function, no congestive heart failure, and mild upper limb hypertension, intervention is indicated beyond 3–6 months of age (class I).
- III. In patients with no hypertension, no heart failure, and normal ventricular function, intervention is indicated at 1–2 years of age (class I), or later whenever the patient presents.

Mode of intervention

- I. For neonatal presentation, surgery is the preferred mode of intervention. Aortic arch hypoplasia, if associated, should also be repaired.
- II. For critically ill neonates who are considered high risk for surgery (shock-like syndrome and severe left ventricular dysfunction), balloon angioplasty may be performed to tide over the crisis (class IIa).
- III. For infants with native coarctation, surgery (class I) or balloon angioplasty (class IIa) may be performed.
- IV. For infants with recoarctation, balloon angioplasty (class I) is the preferred mode of intervention.
- V. For children weighing < 25 kg with native coarctation, balloon angioplasty (class I) or surgery (class IIa), may be performed.
- VI. For children weighing < 25 kg with recoarctation, balloon angioplasty \pm stenting (class I) is the preferred mode of intervention.
- VII. For children weighing > 25 kg and adults, with native coarctation, catheter-based stenting (class IIa) is the preferred mode of intervention.
- VIII. For children weighing > 25 kg and adults, with recoarctation, catheter-based stenting (class I) is the preferred mode of intervention.

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

The following are the indications:

- I. Native coarctation where risk of rupture of the aorta is high (BAV with ascending aorta dilation, nearly atretic isthmus [< 3 -mm diameter], Turner syndrome, age > 60 years, Marfan syndrome).
- II. Recoarctation with aneurysm or pseudoaneurysm at the site of CoA.

Important determinants of long-term prognosis

Residual or recurring coarctation, status of the 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 are important determinants of long-term prognosis.

Follow-up recommendations

- I. Lifelong follow-up is required.
- II. Annual follow-up is required initially, later every 2–3 years if there are no residual lesions.
- III. Clinical assessment should include measuring upper and lower limb blood pressure. Echocardiography should 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, echocardiography alone may not be sufficient for evaluation. cMRI or CTA is recommended every 3–5 years or earlier. cMRI is preferable in postsurgical and post-balloon angioplasty patients whereas CTA is preferred after endovascular stenting.

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

5.7. 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 (subvalvar, 15%, mostly due to discrete membrane) or above the valve (supravalvar, least common). AS is more common in males. BAV has been identified in 1% of the general population; however, the incidence of valvar AS is 0.2–0.4/1000 live births.⁶ BAV occurs in 9% of asymptomatic first-degree relatives of patients with BAV.²¹ Severity of AS usually progresses in 89% of children younger than 2 years, and 61% of children older than 2 years.²² 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.²³

Diagnostic workup

The workup includes clinical assessment, X-ray of the chest, ECG, and echocardiography. It is reasonable to screen first-degree relatives of patients with BAV or unicuspid aortic valve with echocardiography for valve disease and aortopathy. CTA/cMRI may be required in older patients with BAV to assess severity of aortopathy and in select cases of supravalvar AS. Cardiac catheterization is performed primarily for therapeutic balloon valvuloplasty for valvar AS. Exercise testing may be performed for asymptomatic patients with borderline gradients and a normal ECG.

Indications and timing of treatment

Valvar AS

- I. Immediate intervention is required in the following situations:
 - a. Newborns with severe AS who are duct dependent (balloon dilation or surgical valvotomy) (class I).
 - b. Patients with left ventricular dysfunction due to severe AS, regardless of the valve gradient (class I).
- II. Elective balloon dilation is required in the following situations:
 - a. Asymptomatic or symptomatic patients with AS having a peak gradient of >64 mmHg or a mean gradient of >40 mmHg on echo Doppler (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 peak to peak gradient (invasively measured) of ≥ 40 mmHg (class I).
 - c. An 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. Balloon dilation should not be performed for AS in the presence of preexisting AR of more than mild severity (class III).

Subvalvar AS due to discrete membrane. Surgical intervention is indicated in the following situations:

- 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. 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).

Supravalvar AS. Surgical intervention is indicated in the following situations:

- I. Symptomatic patients with a peak instantaneous gradient ≥ 64 mmHg and/or mean gradient ≥ 50 mmHg on echo-Doppler (class I).
- II. Patients with a mean Doppler gradient <50 mmHg, if they have symptoms (exertional dyspnea, angina, syncope), or left ventricular systolic dysfunction, or severe left ventricular hypertrophy attributable to obstruction, or evidence of myocardial ischemia due to coronary ostial involvement (class I).
- III. Asymptomatic patients with a 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 oral hygiene.

Important determinants of long-term prognosis

Residual or recurring stenosis, progressive aortic dilation, and complications related to prosthetic valve function, such as stuck valve leaflet, paravalvular leak, patient-prosthetic mismatch, and pannus formation.

Recommendations for follow-up

- I. All patients with AS require life-long follow-up, irrespective of the type of intervention.
- II. Clinical assessment, ECG, and echocardiography are required, the interval depending on the severity of stenosis.

- III. For 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 conducted annually.
- IV. Echocardiography is the mainstay for follow-up for assessment of aortic valve, ventricular function, and aforementioned 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 a 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.

5.8. Pulmonic stenosis

Background

Pulmonary stenosis (PS) constitutes 8–10% of all patients with CHD.^{6,9} The obstruction is at the valve level in 80–90% of patients. The pulmonary valve is dysplastic in 10–20% of patients.²⁴ Older patients with valvar PS are often asymptomatic, and the diagnosis is made on incidental detection of a murmur. Occasionally, however, they present in heart failure due to right ventricular dysfunction secondary to severe PS. PS may remain stable, progress, or rarely improve. The natural history of patients with valvar PS is excellent with the 1-year, 2-year, and 15-year actuarial survival rate of 97%, 96%, and 94%, respectively.²⁵

Diagnostic workup

The workup includes clinical assessment, X-ray of the chest, ECG, and echocardiography. Cardiac catheterization is performed for balloon valvuloplasty.

Indications and timing of treatment for valvar pulmonic stenosis

- I. Immediate intervention is required in the following situations:
 - 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 is required in the following situations:
 - a. Asymptomatic or symptomatic patients with valvar PS having a peak instantaneous gradient of >64 mmHg on echo-Doppler (class I).
 - b. Neonates and infants with any degree of PS who have hypoxia due to mild hypoplasia of the right ventricle (RV), even if right ventricular function is normal (class IIa).
 - c. Patients with valvar PS due to dysplastic valve, who meet the aforementioned criteria (class IIa).

Mode of intervention

- I. Balloon dilatation (class I) is the preferred mode of intervention.
- II. Surgical intervention is reserved for subvalvar or supra-valvar PS (indications same as in valvar PS), Noonan syndrome (dysplastic valve) with hypoplastic annulus, and failed balloon dilatation (class I).

Recommendations for follow-up

- I. All patients with PS require life-long follow-up.

- II. Clinical assessment, ECG, and echocardiography is 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 up 3-monthly until one year of age. Thereafter, they should be followed up every 1–2 years until 10 years of age and later every 3–5 years. Those with more than mild stenosis (native or after balloon dilation) may be followed up 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.

5.9. Tetralogy of Fallot

Background

Tetralogy of Fallot (TOF) is the commonest cyanotic CHD with a prevalence of 0.4 per 1000 live births, constituting about 5% of all congenital heart defects.⁹ The clinical signs and symptoms generally vary in accordance with the degree of right ventricular outflow tract obstruction. Intermittent hypercyanotic spells are one of the defining features of TOF. Peak incidence of these episodes occurs between the second and sixth month of life, and these become infrequent after 2 years of age.²⁶ Patients with untreated TOF have an estimated 1-year, 3-year, and 10-year survival of 66%, 49%, and 24%, respectively.²⁷

Diagnostic workup

The workup includes clinical assessment, pulse oximetry, ECG, X-ray of the chest, and echocardiography. CTA or cardiac catheterization and angiography should be performed for older children and adults to confirm echocardiographic findings and to delineate coronary artery anatomy and aortopulmonary collaterals.

Indications and timing of surgery

- I. All patients need surgical repair.
- II. In stable patients with minimal cyanosis, total repair is indicated at 6–12 months of age or earlier according to the institutional policy (class I). Those presenting later in life should undergo surgical repair whenever diagnosed.
- III. For symptomatic children aged <6 months 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 is indicated depending on the anatomy and center's experience (class I).
- IV. Lower threshold for earlier surgery if no requirement of the transannular patch is anticipated.
- V. For patients having TOF with absent pulmonary valve who are stable, medical management is indicated until 1 year of age followed by total correction with repair of PA branch dilation/aneurysm (class I).
- VI. For patients with an anomalous left anterior descending artery from the right coronary artery crossing the right ventricular outflow tract, who are likely to need right ventricle to pulmonary artery conduit (class I), the intervention varies as per weight of the patient:
 - a. For those weighing <10 kg and with significant cyanosis, aorto-pulmonary shunt is indicated.
 - b. For those weighing >10 kg, total repair using a conduit, or double-barrel approach after two years of age is indicated.

Important determinants of long-term prognosis

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

Recommendations for follow-up

- I. All patients operated for TOF require life-long follow-up in view of the aforementioned postoperative issues.
- II. Asymptomatic patients with no residual lesion but with free pulmonary regurgitation, not requiring intervention, should be followed up every 1–2 years.
- III. Clinical assessment, ECG, and echocardiography 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 the PA branch for stenosis.
- V. cMRI is an important investigation for follow-up of these patients. In asymptomatic patients, the 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 useful to determine the timing of pulmonary valve replacement.
- VI. Infective endocarditis prophylaxis is indicated for uncorrected patients, patients with percutaneous or surgical pulmonary valve replacement, and after surgical repair, for 6 months. However, all patients with TOF are advised to maintain good oro-dental hygiene even after 6 months of surgical repair.

Indications for pulmonary valve replacement²⁸

The following are the indications:

- I. Symptomatic patients with symptoms attributed to severe right ventricular volume overload with moderate or severe pulmonary regurgitation (class I).
- II. Asymptomatic patients with two or more of the 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/m², right ventricular end-systolic volume >80 ml/m², or right ventricular end-diastolic volume \geq twice the left ventricular end-diastolic volume.
 - c. Right ventricular systolic pressure \geq two-thirds 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).

5.10. VSD with pulmonary atresia

Background

VSD with pulmonary atresia (VSD-PA) is the most severe form of TOF, accounting for 20% of all forms of TOF.²⁹ This CHD usually presents in the neonatal period with cyanosis owing to the right-to-left shunt at the 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. Untreated patients with VSD-PA have a very dismal outcome with the 1-year and 10-year survival being 50% and 8%, respectively.²⁷

Diagnostic workup

The workup includes clinical assessment, pulse oximetry, ECG, X-ray of the chest, and echocardiography. Additional imaging, in the form of cardiac catheterization, CTA/cMRI, or a combination of these, is essential for planning definitive repair.

Indications and timing of intervention

Management depends on the type of VSD-PA, the institutional experience, and the clinical presentation. Generally, this lesion requires a multistage management.

Type A (short-segment VSD-PA with PDA)

- I. In case of presentation with significant cyanosis at less than one year of age: Aortopulmonary shunt (class I) or PDA stenting (class IIa) is indicated depending on the institutional preference and feasibility.
- II. After the 1st intervention or for those presenting at \geq one year of age, total correction is indicated at about 1 year of age because a RV-to-PA conduit is not required (class I).

Type B (long-segment pulmonary atresia with PDA)

- I. In case of those presenting with significant cyanosis at less than one year of age: Aortopulmonary shunt (class I) or PDA stenting (class IIa) is indicated depending on the institutional preference and feasibility.
- II. After the 1st intervention or in those presenting at \geq one year of age (class I), the intervention varies as follows:
 - a. In case of optimal pulmonary blood flow with good-sized PAs, total repair is indicated with RV-to-PA conduit at the age of 3–4 years.
 - b. In case of suboptimal pulmonary blood flow with small-sized PAs, an additional shunt followed by total repair with RV-to-PA conduit is indicated at the age of 3–4 years.
 - c. In case of increased pulmonary blood flow with large-sized PAs, total repair with the RV-to-PA conduit is indicated by the age of 1 year.

Type C (long-segment pulmonary atresia with confluent branch pulmonary arteries supplied by multiple aorto-pulmonary collateral arteries (MAPCAs)) (class I)

- I. In case of neonatal presentation, options are aortopulmonary shunt +/- unifocalisation of MAPCAs, or RV to PA conduit keeping VSD open.
- II. After the 1st intervention or in late presenters, the intervention varies as follows:
 - a. In case of optimal pulmonary blood flow with good-sized PAs, total repair with RV to PA conduit and VSD closure is indicated at 3–4 years of age.
 - b. In case of borderline PAs with large MAPCAs:
 - i. Unifocalisation + RV-to-PA conduit at the age of 6–12 months.

- ii. Total repair with RV-to-PA conduit and VSD closure at the age of 3–4 years.
- c. In case of increased pulmonary blood flow and large PAs, single-stage repair (unifocalisation of MAPCAs + RV-to-PA conduit + VSD closure) is indicated at about 1 year of age preferably with a weight of >10 kg.

Type D (long-segment pulmonary atresia with nonconfluent branch pulmonary arteries supplied by MAPCAs) (class IIa)

- I. In case of neonatal presentation, aortopulmonary shunt + unifocalisation of MAPCAs is indicated.
- II. After the 1st intervention or at late presentation:
 - a. Unifocalisation + RV-to-PA conduit at the age of 6–12 months.
 - b. Total repair with RV-to-PA conduit and VSD closure at the age of 3–4 years.

Important determinants of long-term prognosis

These include conduit obstruction, degeneration, PA branch stenosis, residual VSD, aortic root dilatation, and functional abnormalities such as 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-PA require life-long follow-up.
- II. Annual follow-up is required for 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. cMRI is an important investigation for follow-up of these patients to determine the timing of conduit revision and pulmonary valve replacement.
- V. Infective endocarditis prophylaxis is indicated in non-corrected or palliated patients with cyanosis; in patients with conduits; in patients who have undergone pulmonary valve replacement; and after surgical repair for 6 months.

All patients are advised to maintain good oro-dental hygiene even after 6 months of surgical repair.

Indications for conduit replacement³⁰

The following are the indications:

- I. Symptomatic patients with right ventricular pressure greater than 80 mmHg on echo Doppler and/or moderate to severe pulmonary regurgitation (class I).
- II. Asymptomatic patients with right ventricular pressure greater than 80 mmHg on echo Doppler with at least one of the following symptoms (class IIa):
 - a. Depressed right ventricular function
 - b. Progressive right ventricular dilation
 - c. Progressive tricuspid regurgitation
 - d. Sustained atrial or ventricular arrhythmias

5.11. Transposition of the great arteries

Background

Transposition of the great arteries (TGA) is the most common cyanotic CHD at birth, accounting for approximately 5% of all CHDs.⁶ In 70% of cases, there is no associated defect (simple TGA) apart from ASD, PDA, or insignificant VSD. Association of TGA with other defects such as large VSD, left ventricular outflow tract obstruction or coarctation, etc., is referred to as complex TGA.³¹ 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 one year being close to 90%.³² With the advent of improved surgical techniques and postoperative care, long-term survival is more than 90% with very low reintervention rates.³³

Diagnostic workup

The workup includes clinical assessment, pulse oximetry, ECG, X-ray of the chest, and echocardiography. Echocardiography is the key tool to establish the diagnosis and to assess site and adequacy of intermixing, associated malformations, coronary artery anatomy, and adequacy of the left ventricle to support systemic circulation after an arterial switch operation (ASO). Cardiac catheterization is generally performed for balloon atrial septostomy. CTA/cMRI is rarely required to clarify anatomy of the aortic arch or to evaluate 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 in neonates is achieved by intravenous infusion of prostaglandin E1 to keep the ductus arteriosus open, and/or balloon atrial septostomy to create an adequate ASD (class I).

Timing and type of surgery

- I. For patients with TGA with intact ventricular septum presenting soon after birth, arterial switch operation (ASO) is the best option (class I). ASO should be done between 7 days to 3 weeks of age; earlier if the baby is unstable or has associated persistent pulmonary hypertension of the newborn. Exact timing is based on institutional preference but is best done before 4 weeks.
- II. For patients with TGA with intact ventricular septum presenting beyond 3–4 weeks of life with regressed left ventricle, the type of surgery varies as follows:
 - a. If the patient is presenting between the age of 1 and 2 months, ASO is indicated; extracorporeal membrane oxygenator (ECMO) support may be required in some cases (class IIa).
 - b. If the patient is presenting between the age of 2 and 6 months, the options for surgical repair are: ASO with ECMO support or rapid two-stage ASO^{ac} or an atrial switch operation (if rapid two-stage or ECMO is not feasible) (class IIa).
 - c. If the patient is presenting between the age of 6 months and 2 years, atrial switch operation (Senning or Mustard operation) (class IIa) is indicated. Rapid two-stage ASO^{ac}

^{ac} The first stage of rapid two-stage ASO involves retraining of the regressed left ventricle by performing PA banding along with the addition of a modified aortopulmonary shunt as the first stage.

may be considered in select cases after detailed evaluation (class IIb).

- d. If the patient is presenting later in life with a regressed left ventricle, the patient must be assessed for operability; atrial switch operation is recommended in operable patients.
- III. For patients with TGA with a large VSD and/or a large PDA, ASO with VSD and/or PDA closure by 6 weeks of age (class I) is recommended. These patients tend to develop early pulmonary vascular disease and may become inoperable by the age of 6 months to one year.
- IV. For patients with TGA with VSD and CoA, ASO with VSD closure and arch repair is recommended as soon as possible (class I). It is preferable to repair all lesions in a single stage.
- V. For patients with TGA with VSD and significant left ventricular outflow tract obstruction, the type and timing of surgery varies as follows (class I):
 - a. In those with subvalvar pulmonary obstruction with normal or near-normal pulmonary valve and pulmonary annulus, ASO with resection of subvalvar stenosis is recommended.
 - b. If the obstruction involves the pulmonary valve or is subpulmonary but not amenable to resection, the approach is as follows:
 - i. For neonates and infants presenting with significant cyanosis, surgical options depend on the patient's age and surgeon's preference:
 - a) Systemic-to-pulmonary artery shunt (at any age) followed by Rastelli-type repair (at 2–3 years of age, or when the child weighs >10 kg), or root translocation later.
 - b) Réparation à l'Etage Ventriculaire procedure (usually done at the age of 4–6 months)
 - c) Pulmonary root translocation (usually done at the age of 6–12 months).
 - d) Nikaidoh procedure (usually done beyond 6–9 months of age)
 - ii. For older, stable patients, presenting beyond 2–3 years of age, one of the following surgeries may be opted for: 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) is recommended.

Important determinants of long-term prognosis

Long-term prognosis after surgery depends on the type of surgery performed. Attention should be paid to specific issues such as residual defects, coronary insufficiency with resultant myocardial ischemia/ventricular dysfunction, supravalvar obstruction of outflow tracts, neo-aortic valve regurgitation, neo-aortic 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 until the onset of adult life, and every 2–3 years thereafter.

- III. Follow-up visits should include clinical assessment, ECG, and echocardiography.
- IV. Holter monitoring is done when suspecting arrhythmias or myocardial ischemia. Frequent Holter monitoring may be required after 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 carried out in cases who had intramural coronary artery or difficult coronary transfer at the time of surgery.

Infective endocarditis prophylaxis is recommended in patients with cyanosis, in cases with use of conduits or other prosthetic material during surgery, and for 6 months after definitive surgery. However, all patients are advised to maintain good oro-dental hygiene even after 6 months of definitive surgery.

5.12. Double outlet right ventricle

Background

Double outlet right ventricle (DORV) constitutes less than 1% of all congenital heart defects.⁶ VSD is the only outlet for the left ventricle, and it could be subaortic (60–65%), subpulmonary (20–25%), doubly committed (3–5%) or remote (noncommitted, 5%).³⁴ The clinical presentation is variable and depends on the location of VSD, the presence or absence of obstruction to pulmonary blood flow, and associated cardiac anomalies.⁹ Clinical presentation of DORV can be divided into three types: TOF-like presentation, VSD-like presentation, and TGA-like presentation. 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

The workup includes clinical presentation, ECG, X-ray of the chest, and echocardiography. Cardiac catheterization is performed in late presenters with pulmonary hypertension suspected of having high PVR. It may also be done in other patients, especially adults, to define the anatomy better. The presence of associated arch hypoplasia or coarctation may necessitate CTA or cMRI. Three-dimensional reconstruction of CT images helps to assess routability of VSD to aorta.

Indications and timing of surgery

Surgery is indicated for all patients with DORV except in those with irreversible pulmonary vascular disease. Timing and the type of surgery depends on the DORV variant (class I):

- I. For those with DORV with subaortic VSD and severe PS (TOF-type DORV), the type of surgery varies as follows:
 - a. For those presenting with significant cyanosis at the age of <3–4 months: aortopulmonary shunt is indicated.
 - b. For those presenting with significant cyanosis at the age of >3–4 months, total repair with closure of VSD and infundibular resection is indicated.
 - c. For stable patients with no or minimal cyanosis, total repair with closure of VSD and infundibular resection is indicated by the age of 6–12 months. or later if presenting beyond this age.
- II. For those with DORV and large subaortic VSD and pulmonary hypertension (VSD-type DORV), VSD closure is indicated by 6 months of age. In patients presenting beyond 6 months of age, VSD can be closed, if operable on assessment.

- III. For those with DORV and subpulmonary VSD and pulmonary hypertension (TGA-type DORV), ASO with VSD closure is indicated by 6 weeks of age. If presenting beyond 3 months, patient should be evaluated for operability.
- IV. For those with DORV with subpulmonary VSD and severe PS, the type of surgery varies as follows:
 - a. If pulmonary obstruction is localized, for example, subvalvar fibrous membrane or ridge, ASO with resection of subvalvar stenosis is indicated.
 - b. If pulmonary obstruction is tubular or valvar, one should try to perform biventricular repair as far as possible, and this may necessitate a conduit from the RV to PA. Univentricular palliation is done if biventricular repair is not possible. A systemic-to-PA shunt may be required 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. For those with DORV and remote VSD or associated with other complex anatomy, biventricular or univentricular repair is indicated depending on the anatomy.

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 there is no residual defect after surgery, annual follow-up is needed until adult life and then every 2 years.
- III. Follow-up visits should include clinical assessment, ECG, and echocardiography.
- IV. Holter monitoring should be done every 2–3 years after the age of 5 years.

Infective endocarditis prophylaxis is recommended for patients with cyanosis and for 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.

5.13. Congenitally corrected TGA

Background

Congenitally corrected TGA (ccTGA) is a congenital heart defect characterized by atrio-ventricular and ventriculo-arterial discordance. This double discordance results in physiologically corrected circulation as the great arteries receive appropriate blood. ccTGA is a rare condition occurring in 0.3 per 10,000 live births and constitutes 0.5% of all congenital heart defects.⁶ Although the physiology is corrected, anatomy is not corrected, and the morphologic RV supports systemic arterial circulation. In more than 90% of cases, ccTGA is accompanied by other cardiac lesions: VSDs in 80%, PS (obstruction of the outflow from 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%.³⁵ Mismatch between visceral situs and cardiac position is common, with dextrocardia/mesocardia in one-fourth of patients. The risk of developing CHB is 2% per year, and about 30% patients develop CHB by adulthood.³⁶ The function of the RV deteriorates after the second decade even in those without any associated anomaly.³⁷

Diagnostic workup

The workup includes clinical assessment, ECG, X-ray of the chest, and echocardiography. Cardiac catheterization may have to be performed for demonstrating coronary anatomy or for measurement of PA pressure and PVR.

Indications and timing of surgery

The indications and timing of surgery in ccTGA patients depends on the presence of associated anomalies.^{38,39}

General recommendations

- I. Tricuspid valve (systemic atrioventricular 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 the left ventricle is functioning at systemic pressure and when such surgery is feasible (class IIa).

Indications and timing for specific groups of ccTGA

- I. In case of no associated anomalies, patient should be regularly followed to look for development of tricuspid regurgitation, CHB, or right ventricular dysfunction (class I). Neonatal double switch operation may be considered (class IIb).
- II. If associated with large VSD, the type of surgery varies as per age:
 - a. At the age of <3 months, PA banding is indicated, followed later by double switch operation (atrial plus arterial switch) (class I).
 - b. At the age of >6 months, double switch operation (atrial plus arterial switch) is indicated, provided that the patient has not developed irreversible pulmonary vascular disease (class I).
 - c. At the age of 3–6 months, PA banding followed by double switch operation or direct double switch operation can be done, depending on institutional policy (class IIa).
- III. If associated with large VSD and severe left ventricular outflow tract obstruction (PS), the type of surgery varies as follows:
 - a. If VSD is routable, double switch operation (atrial switch plus Rastelli) (class I) is indicated; univentricular repair pathway may be followed if the surgeon is not comfortable doing double switch operation and saturation is low (class IIa).
 - b. If VSD is not routable, the type of intervention varies as follows:
 - i. If saturation is good, patient may be regularly followed after informed discussion with the family (class IIa).
 - ii. If saturation is low, univentricular repair pathway (class I) is indicated.
- IV. If associated with CHB, permanent, dual chamber pacemaker implantation (class I) is indicated.
- V. If associated with severe tricuspid regurgitation in an adult, cardiac catheterization should be done to measure the pressure in the left ventricle.
 - a. In case of good right ventricular function and prepared left ventricle, double switch operation and tricuspid valve repair (class IIa) is indicated.
 - b. In case of good right ventricular function and low left ventricular pressure, tricuspid valve repair/replacement (class IIb) is indicated.

- VI. If associated with severe right ventricular dysfunction in an adult, possible management options are: PA banding/cardiac resynchronization therapy/cardiac transplant (class IIa).

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 testing, and electrophysiological study may be indicated in select patients.

Infective endocarditis prophylaxis is recommended for all patients with cyanosis and in cases with conduits or other prosthetic material in the heart. However, all patients with ccTGA are advised to maintain good oro-dental hygiene.

5.14. Univentricular hearts (single ventricles)

Background

Univentricular hearts are defined as presence of one ventricle instead of two, or the second ventricle is rudimentary without an inlet portion. It also includes those congenital heart defects where two-ventricle (or biventricular) repair is not possible. Classical examples include double inlet left ventricle, tricuspid atresia, hypoplastic left heart syndrome, unbalanced AVSD, and nonroutable or multiple VSDs. These defects are grouped together as “functional univentricular” heart because the management is on the lines of single ventricle.⁴⁰ Univentricular hearts constitute 2.8% of all congenital heart defects.^{9,41} The clinical presentation depends on the ratio of pulmonary to systemic blood flow.

Diagnostic workup

The workup includes clinical assessment, pulse oximetry, ECG, X-ray of the chest, and echocardiography. Cardiac catheterization is required for assessing PVR in patients with unrestricted pulmonary blood flow, those presenting late, and for interventions such as occlusion of aortopulmonary collaterals and in certain operated patients with treatable complications. cMRI may be required for older children and adults who have undergone Fontan operation for assessing ventricular volumes and function.

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.³⁹ The treating physician must inform and discuss the details with the family before surgery.

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

- I. For those presenting in the neonatal period or within 2–3 months of life, the type of intervention is as follows (class I):
 - a. In those with increased pulmonary blood flow, PA banding is indicated at 4–6 weeks of age, preferably before the age of 3 months.
 - b. In those with decreased pulmonary blood flow (PS group): systemic-to-pulmonary artery shunt or stenting of the ductus arteriosus (depends on institutional policy and clinical scenario) is done if systemic arterial saturation is consistently

below 70–75% or if there is pulmonary atresia with duct-dependent pulmonary circulation.

- II. For those presenting later in life or who have undergone first surgery earlier, the type of intervention varies as follows:
 - a. In those with pulmonary hypertension and no PS, most patients presenting beyond the age of 3–4 months would be unsuitable for PA banding or any definitive repair in the future owing to irreversible increase in PVR. A minority may continue to have low resistance and should be offered PA banding after complete evaluation (including cardiac catheterization for operability).
 - b. In those with normal pulmonary pressure and resistance due to PS/previous PA banding/previous aortopulmonary shunt, the type of intervention varies as follows:
 - i. Bidirectional Glenn procedure is indicated between 6–12 months of age (class I).
 - ii. Total cavopulmonary connection or completion of Fontan procedure (preferably extracardiac) is indicated between 4 and 7 years of age when the child weighs 15–20 kg. If Fontan is required earlier (owing to increasing cyanosis or due to pulmonary arteriovenous malformations), a lateral tunnel Fontan can be performed at an age of 3–4 years. Fenestration of the Fontan circuit is indicated in high-risk cases.
 - iii. In adults who have a balanced pulmonary blood flow and are asymptomatic with minimal desaturation, the role of palliative surgeries is not clear; however, such patients should remain under follow-up.

Important determinants of long-term prognosis

The patients palliated for univentricular hearts experience a number of morbidities in the long term. These include development of ventricular dysfunction, growth failure, worsening cyanosis, arrhythmias, atrioventricular 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 it should be at least once a year in stable cases.
- II. Follow-up includes clinical assessment, oxygen saturation, ECG, X-ray of the chest, and echocardiography. In addition, hematological, kidney, and liver function tests should be done for all post Fontan patients. cMRI may be performed at least once in adult life and then repeated, if indicated. Periodic imaging of the liver by ultrasound, and additional imaging by CT/MRI/biopsy is indicated, starting 10 years after total cavopulmonary connection.⁴²
- III. After surgery, aspirin (3–5 mg/kg/day) is recommended for all patients. Oral anticoagulants (warfarin) and sildenafil are recommended in a select group or as per institution policy.⁴³
- IV. 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.

Infective endocarditis prophylaxis is recommended in patients with cyanosis and in cases with conduits or other prosthetic material in the heart. However, all patients with univentricular heart are advised to maintain good oro-dental hygiene.

5.15. Persistent truncus arteriosus

Background

Persistent truncus arteriosus accounts for less than 1% of all congenital heart defects. Aortic arch interruption or CoA is found in 15–20% of patients with truncus arteriosus. 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.⁴⁴

Classification of truncus arteriosus⁴⁵

The classification is as follows:

- I. Type A1: The aorta and main PA originate from a single large common trunk.
- II. Type A2: Both pulmonary arteries arise separately and directly from the truncus.
- III. Type A3: One PA 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

The workup includes clinical assessment, pulse oximetry, X-ray of the chest, ECG, and echocardiography. CTA and cMRI are useful when the anatomy is unclear on echocardiography, especially for evaluation of the aortic arch. Cardiac catheterization is indicated in patients presenting late for operability assessment.

Ideal age for surgery

Surgery is indicated for all, unless the patient is inoperable due to irreversible pulmonary vascular disease.

- I. In case of uncontrolled heart failure, surgical repair is indicated as soon as possible (class I).
- II. In case of controlled heart failure, surgical repair is indicated by 3–6 weeks of age (class I).

Type of surgery

Total repair is indicated using the RV-to-PA conduit. The prospects of repeat surgeries in future for conduit obstruction should be discussed with the family. Truncal valve repair is performed if the valve is regurgitant.

Contraindication for surgery

Severe pulmonary arterial hypertension with irreversible pulmonary vascular occlusive disease (class III) is a contraindication for surgery. Signs of inoperability include age more than 1 year, resting systemic arterial saturation <85%, and absence of cardiomegaly.

Important determinants of long-term prognosis

These include residual or progressive pulmonary hypertension, the 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 aforementioned post-operative issues.

- II. Follow-up after surgery includes clinical assessment, X-ray of the chest, ECG, and echocardiography at 1, 6, and 12 months and yearly thereafter in stable cases.

Infective endocarditis prophylaxis is recommended after surgical repair due to the presence of conduit. All patients are advised to maintain good oro-dental hygiene.

5.16. Total anomalous pulmonary venous connection

Background

Total anomalous pulmonary venous connection (TAPVC) accounts for 1% of all patients born with congenital heart defects.⁹ TAPVC is classified into four types depending on the site of drainage: supracardiac (45–50%), cardiac (15–20%), infracardiac (26–28%), and mixed (where the drainage is at two or more sites, 5–8%).⁴⁶ Each type can be obstructive or nonobstructive. 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 first year of life. Exceptionally, some may present later during adult life with features suggestive of a large ASD and mild desaturation.

Diagnostic workup

The workup includes clinical assessment, ECG, X-ray of the chest, and echocardiography. Cardiac catheterization may be required for older children and adults for assessing PA pressures and defining the anatomy.

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 for those with nonobstructive TAPVC, even if they are asymptomatic.
- IV. Those presenting late should be evaluated for the onset of pulmonary vascular disease and operated if the data suggests operable status.

Important determinants of long-term prognosis

These include pulmonary vein stenosis, pulmonary hypertension, stenosis of surgically created anastomosis, and late-onset arrhythmias.

Recommendations for follow-up

- I. After surgery, patients should be followed up at one month, 6 months, and then annually for 5 years if there is no residual defect and pulmonary hypertension.
- II. ECG and echocardiography should be carried out at each visit.
- III. CTA/cMRI should be carried out in operated patients suspected to have developed 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 later.

Infective endocarditis prophylaxis is indicated for noncorrected patients and for patients after surgical repair for 6 months. However, all patients with TAPVC are advised to maintain good oro-dental hygiene after this period also.

5.17. Ebstein's anomaly of the tricuspid valve

Background

Ebstein's anomaly is rare, constituting less than 1% of all congenital heart defects.⁶ Patients usually present either in the neonatal period or during adolescence or adult life. Older patients present with murmur, arrhythmias, or cyanosis. The estimated survival is 76% and 53% at 10-year and 15-year follow-up, respectively, when diagnosed in adulthood.⁴⁷ Prognosis is poor in those diagnosed during fetal or neonatal life.

Diagnostic workup

The workup includes clinical assessment, ECG, X-ray of the chest, and echocardiography. Cardiac catheterization is rarely performed and may be done for evaluating coronary arteries in older patients (age >40 years) planned for surgical repair of Ebstein's anomaly. cMRI provides quantitative measurement of the functional right ventricular size, volume, and function, which are important when planning surgical repair.

Indications and timing for treatment

Tricuspid valve repair is best done at about 2 years of age for stable cases.

- I. Surgery is indicated (class I) for those with any of the following: deteriorating exercise capacity, cyanosis (oxygen saturation <90%), paradoxical embolism, progressive cardiomegaly on chest X-ray (CT ratio >0.65), or progressive dilation or dysfunction of the RV on echocardiography.
- II. If the patient is symptomatic with arrhythmias, catheter ablation is indicated. Surgery is indicated, if arrhythmia is not amenable to catheter ablation (class IIa).

Types of surgery

The type of surgery depends on the underlying anatomy and size of the functional ventricle and is as follows:

- 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 of the chest, and echocardiography should be performed at each visit. Holter monitoring, exercise testing, and cMRI may be required for select patients.
- III. Asymptomatic patients who are not candidates for surgery can be followed up every 2–3 years.
- IV. Those who have undergone tricuspid valve replacement with a prosthetic valve should be closely monitored by INR testing for optimal anticoagulation.

Infective endocarditis prophylaxis is indicated for 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.

5.18. Mitral regurgitation and aortic regurgitation

Background

Mitral regurgitation (MR) and aortic regurgitation (AR) occur most commonly secondary to acute or chronic rheumatic heart disease, and both may coexist in some patients. 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 infective endocarditis. 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 both, chronic MR and chronic AR.

Diagnostic workup

The workup includes clinical assessment, ECG, X-ray of the chest, and echocardiography.

Measurement of left ventricular dimensions by serial echo-Doppler helps in deciding the timing of valve surgery. Exercise testing is performed for select cases where symptoms are out of proportion to severity of the valve lesion. Cardiac catheterization is rarely required. CTA or cMRI may be needed in select cases of AR to define the ascending aorta and aortic arch.

Role of drug therapy

- I. Drugs are not 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 a short term prior to surgery but may be used for a long term in patients with symptoms, or patients with left ventricular dysfunction who are not candidates for valve surgery. It may be used in asymptomatic patients with normal left ventricular systolic function to extend the compensated phase, prior to 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 treatment of acute MR; invasive blood pressure (BP) monitoring is required in these cases.
- V. Anticoagulants (oral) to be prescribed 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^{48,49}

- 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. For asymptomatic patients with severe MR, surgery is indicated if any of the following is 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 >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 regurgitation⁴⁸

- I. Symptomatic patients with moderate to severe AR (class I)
- II. For asymptomatic patients with severe AR, surgery is indicated if any of the following is 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 surgery⁵⁰

- I. Valve repairs are preferable to valve replacements (class I).
- II. Valve replacement in those in whom the valve cannot be repaired (class IIa).
 - a. The Ross procedure is indicated for young patients with nonrheumatic AR (if expertise is available).
 - b. The bioprosthetic valve is indicated in the following cases:
 - i. Female patients planning pregnancy in future
 - ii. Compliance with oral anticoagulation is dubious
 - c. Prosthetic metallic valve replacements are indicated for the rest of the patients.

Anticoagulation after valve surgery⁵¹

- I. Oral anticoagulant drugs such as warfarin or other anticoumarin drugs are to be used.
 - a. The desired INR after surgery should be as follows:
 - i. 3.0 (\pm 0.5) after mitral valve replacement
 - ii. 2.5 (\pm 0.5) after aortic valve replacement
 - iii. 2.5 (\pm 0.5) after valve repair, bioprosthetic valve
 - b. Patients taking warfarin should be educated about the importance of maintaining INR in the therapeutic range; the effect of diet, medicines, etc. on the INR; 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 the patient is on oral anticoagulant drugs. Dental surgery is safe with therapeutic levels of the INR.
 - c. Duration of anticoagulation after surgery is as follows:
 - 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) should be given in addition to anticoagulation (class I) for 6 months after valve repair and bioprosthetic valve implantation, and lifelong in case of prosthetic metallic valves.

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. For asymptomatic patients with MR or AR, clinical assessment, ECG, and echocardiography are recommended as per the following frequency:
 - a. For patients with mild MR or AR, clinical assessment is recommended every year and echocardiography every 2 years.
 - b. For patients with moderate MR or AR, clinical assessment is recommended every 6 months and echocardiography every year.
 - c. For patients with severe MR or AR, clinical assessment and echocardiography is recommended every 6 months.

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

- III. For operated patients with no residual abnormality: clinical assessment, ECG, and echocardiography are recommended as follows:
 - a. For patients with bioprosthetic valve or after valve repair, follow-up is recommended every 1–2 years.
 - b. For patients with prosthetic metallic valves, follow-up is recommended every year. In addition, these patients require frequent monitoring of INR and fluoroscopy (for valve motion).
- IV. Postsurgical patients with residual valve abnormality are followed up as for native valve regurgitation.

Infective endocarditis prophylaxis

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, the peri-apical region of teeth, or perforation of the oral mucosa, in patients with prosthetic heart valves, patients with use of prosthetic material for cardiac valve repair, such as annuloplasty rings and chords and patients with previous infective endocarditis (class IIa).⁵¹

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Conflicts of interest

The authors have none to declare.

Disclaimer

These recommendations are for use by the physicians only and are not to be used for medicolegal purposes.

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