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. Abridged Secondary Publication

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Justification: 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 10th and 11th of August 2018 at the All India Institute of Medical Sciences, New Delhi. The meeting was supported by Children's HeartLink, a non-governmental organization based in Minnesota, USA. **Objectives:** To frame evidence based guidelines for (i) indications and optimal timing of intervention in common congenital heart diseases; (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 (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 congenital heart diseases (tetralogy of Fallot, transposition of great arteries, univentricular hearts, total anomalous pulmonary venous connection, Ebstein anomaly and others). In addition, protocols for follow-up of post surgical patients are also described, disease wise.

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 of 9/1000, 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 healthcare system. Approximately 10% of infant mortality in India may be accounted for, by CHDs.

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 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 the late presenters have advanced level of pulmonary hypertension, ventricular dysfunction, hypoxia, polycythemia, etc. The outcome after surgery in such patients are expected to be suboptimal with longer 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 management of CHDs in India, including the timing of intervention.

A statement on "consensus on timing of intervention for common congenital heart disease" which 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. In the intervening 10 years, a number of pediatric cardiac centres have been established and overall the numbers of interventions have increased by several folds. Considering the growing population of post-operative patients including those needing regular follow-up, we added guidelines and protocols for follow-up of these patients.

PREAMBLE

1. Every pediatrician/cardiologist/other healthcare provider must strive to get a complete diagnosis on a child suspected of having heart disease, with the help of a higher centre, if needed.
2. The proposed guidelines are meant to assist the health care provider (pediatrician, cardiologist, pediatric cardiologist) in managing cases of congenital heart diseases 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, if 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 favour 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 guidelines and protocols for follow-up of patients who have undergone surgery/catheter interventions for CHD.

GUIDELINES FOR INDIVIDUAL CONGENITAL HEART DEFECTS

Atrial Septal Defect (*ASD*)

Diagnostic work-up: Physical examination, ECG, *X*-ray chest, echocardiography and cardiac catheterisation (in select cases).

Types of Atrial septal defect: Ostium secundum (~75%); Ostium primum (15%-20%); Sinus venosus (5%-10%); and Coronary sinus (<1%).

Patent foramen ovale: Small defect in fossa ovalis region with a flap with no evidence of right heart volume overload. Diagnosed on echocardiography, is a normal finding in newborns.

Indication for closure: 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.

Contraindications for closure: Severe pulmonary arterial hypertension or irreversible pulmonary vascular disease *(Class III)*.

Ideal Age of Closure

Asymptomatic child: 2-4 years *(Class I)*. For sinus venosus defect surgery may be delayed to 4-5 years *(Class IIa)*.

Symptomatic ASD: Rarely seen in infants. Present with congestive heart failure, pulmonary arterial hypertension. Early closure is recommended *(Class I)* after ruling out associated lesions such as left ventricular inflow obstruction, aortopulmonary window, total anomalous pulmonary venous drainage, etc.

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 is within operable range *(Class I)*.

Method of Closure

Surgical: Established mode *(Class I)*.

Device: For secundum ASDs with adequate rims and weight of child >15kg *(Class I)*.

Recommendations for Follow-up

Follow-up after surgical closure: Clinical and echo in the first year only. No further follow-up required if no residual disease, 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.

Follow-up after device closure: (*a*) Anti-platelet agents for total duration of 6 months (*b*) Echocardiography: - At discharge, 1 month, 6 months, 1 year, then every 3-5 years.

IE prophylaxis: It 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 (*VSD*)

Diagnostic Work-up: Physical examination, ECG, X-ray chest, echocardiography and cardiac catheterisation (in select cases).

Classification of Ventricular Septal Defect

Perimembranous: 80%; Outlet or sub-pulmonary (doubly committed): 5%-7%; Inlet: 5%-8%; and muscular: 5%-20%, these could be central (mid muscular), apical, marginal (anterior, septal-free wall area) or multiple, "swiss cheese" type.

Indications and Timing of Closure (All Class I recommendations)

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 patient has an episode of endocarditis or develops cusp prolapse with aortic regurgitation or develops progressive significant right ventricular outflow tract obstruction.

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;

Large VSD: (*a*) Poor growth/congestive heart failure not controlled with medications (furosemide/spironolactone or enalapril +/- digoxin): As soon as possible; (*b*) Controlled heart failure: By 6 months of age.

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

Contraindications for Closure: Severe pulmonary arterial hypertension with irreversible pulmonary vascular disease *(Class III)*.

Method of Closure

Surgery: Conventionally patch closure is done. Pulmonary artery banding to be considered for patients with multiple VSDs, inaccessible VSDs and those with contraindications for cardio-pulmonary bypass.

Device closure: For VSDs with adequate rims around defect and weight of child >8kg.

Recommendations for Follow-up

Follow-up after surgery: Clinical, ECG and echo in the first year only. No further follow-up 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.

Follow-up protocol for device closure: Anti-platelet agents for total duration of 6 months.

IE prophylaxis: It 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 (*AVSD*)

Diagnostic Work-up: Physical examination, ECG, *X*-ray chest, echocardiography and cardiac catheterisation (in select cases).

Types of AVSD

I. *Complete AVSD*: Large septal defect with an atrial component (ostium primum defect) and ventricular component (inlet septal defect), common atrioventricular valve ring and common atrioventricular valve. Generally associated with large left-to-right shunt, pulmonary arterial hypertension and congestive heart failure.

II. *Partial AVSD*: Two separate atrioventricular valves and primum atrial septal defect. Cleft of the anterior leaflet of atrioventricular valve is common with variable degree of regurgitation.

III. *Intermediate AVSD*: Two separate atrioventricular valves with primum atrial septal defect and small restrictive inlet ventricular septal defect.

IV. *Unbalanced AVSD*: One of the ventricles is hypoplastic. This form is usually associated with complex congenital heart defects such as heterotaxy syndrome (isomerism).

Varying degree of atrioventricular valve regurgitation may be associated with AVSD.

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 atrioventricular valve regurgitation may necessitate early surgery in partial or intermediate forms.

IV. Pulmonary artery banding is reserved for complex cases and patients with contraindications for cardio-pulmonary bypass *(Class IIb).*

V. Surgery for moderate to severe left atrioventricular valve regurgitation is recommended as per the guidelines for mitral regurgitation, discussed later *(Class I).*

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.

III. IE prophylaxis 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 (*PDA*)

Diagnostic Work-up: Clinical assessment, *X*-ray chest, ECG, Echocardiography. Cardiac catheterisation is usually performed for device closure.

Ideal Age of Closure

I. Large/moderate PDA (significant left heart volume overload, congestive heart failure, pulmonary arterial hypertension): Early closure (by 3 months) *(Class I).*

II. Moderate PDA (Some degree of left heart overload, mild to moderate pulmonary arterial hypertension, no/mild congestive heart failure): 6 months-1 year *(Class I).* If failure to thrive, closure can be accomplished earlier *(Class IIa).*

III. Small PDA (Minimal or no left heart overload. No pulmonary hypertension or congestive heart failure): Between 12-18 months *(Class I).*

IV. Silent PDA (Diagnosed only on echo Doppler. Hemodynamically insignificant, produce no murmur and there is no pulmonary hypertension): Closure not recommended *(Class III).*

Contraindication for closure: PDA associated with severe pulmonary arterial hypertension with irreversible pulmonary vascular disease, and silent PDA (*Class III).*

Method of Closure: Surgical: Established mode *(Class I)*. Device closure: Preferred for children >6kg as less invasive *(Class I).*

Recommendations for Follow-up

I. Clinical assessment, ECG and echo at one-year post

intervention. No further follow-up required if no residual defect or pulmonary hypertension. Patient/ guardians should be explained about reporting to a hospital in case of any cardiac symptoms.

II. IE prophylaxis 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 2 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).*

Prophylactic Indomethacin or Ibuprofen therapy: Not recommended *(Class III).*

Aortopulmonary Window

Diagnostic Work-up: Clinical assessment, *X*-ray chest, ECG, Echocardiography, Cardiac catheterisation and CT Angiography (select cases).

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).*

***Contraindication for closure*:** Severe pulmonary arterial hypertension with irreversible pulmonary vascular disease *(Class III).*

***Method of Closure*:** Surgical patch repair *(Class I)*, transcatheter device closure in select cases with a restrictive defect.

Recommendations for Follow-up

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

II. IE prophylaxis 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 (*CoA*)

Diagnostic work up: Clinical assessment, X-ray chest, ECG, Echocardiography, CT angiography/cardiac MRI (in select cases when anatomy is unclear, and for follow-up in adults), cardiac catheterisation (if intervention is planned).

Indications for intervention

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

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

III. Patients with gradient <20mmHg 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 <20mmHg 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).*

Mode of Intervention

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

II. Critically ill neonate who are considered high risk for surgery (shock like syndrome, 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 re-coarctation: Balloon angioplasty *(Class I).*

V. Children <25 kg with native coarctation: Balloon angioplasty *(Class I)* or Surgery *(Class IIa).*

VI. Children <25 kg with re-coarctation: 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 re-coarctation: Catheter based stenting *(Class I).*

IX. Elective endovascular stenting of aorta is contra-indicated in children < 10 years of age *(Class III).*

Follow-up Recommendations

I. Lifelong follow-up is required. Annual follow-up initially; later every 2-3 years if no residual lesions. Follow-up should include clinical assessment (upper and lower limb blood pressure) and echocardiography. Beyond 5 years of age, cMRI or CT angiography may be required.

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

III. 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 (*AS*)

Diagnostic Work-up: Clinical assessment, *X*-ray chest, ECG, Echocardiography, CT Angiography/cardiac MRI (in select cases), cardiac catheterisation (primarily for therapeutic balloon valvuloplasty for valvar AS), Exercise test (in select cases).

Indications and Timing of Treatment

Valvar 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 >64mmHg peak or >40mmHg mean or peak to peak gradient of ≥50mmHg, 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 ≥ 40mmHg *(Class I).*

(*c*) Asymptomatic child or adolescent with a peak systolic valve gradient (invasively measured) of ≥ 40mmHg 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).*

Subvalvar AS 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 aortic regurgitation 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).*

V. Intervention not indicated for asymptomatic patients with gradient of < 30 mmHg with no or trivial aortic regurgitation *(Class III).*

Supravalvar AS

Surgical intervention indicated in:

I. Symptomatic patients with peak instantaneous gradient ≥ 64 mmHg and/or mean gradient ≥ 50mmHg 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, 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 ≥50mmHg 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.

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 echo are required; the interval depending on the severity of stenosis.

III. Patients who have significant AS and are planned for an intervention should refrain from any sporting activity. Those with asymptomatic moderate stenosis can participate in low- or moderate-intensity sports. Patients with mild degree of stenosis can participate in all sports.

IV. IE prophylaxis is recommended in patients with a prosthetic valve.

Pulmonic Stenosis (PS)

Diagnostic Work-up: Clinical assessment, X-ray chest, ECG, Echocardiography, cardiac catheterisation and angiography (primarily for therapeutic balloon valvuloplasty), CT Angiography/cardiac MRI (for peripheral pulmonic stenosis).

Indications and Timing of Treatment

Valvar pulmonic stenosis

I. Immediate intervention required for:

(*a*) Newborns with severe PS with duct dependent pulmonary blood flow *(Class I).*

(*b*) Infants or children 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 valvar PS having peak instantaneous gradient by echo-Doppler of >64mmHg *(Class I).*

(*b*) Neonates and infants with any degree of PS who have mild hypoxia due to mild hypoplasia of right ventricle, even if right ventricular function is normal *(Class IIa).*

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

Mode of intervention: Balloon dilatation *(Class I)*; surgical intervention reserved for: subvalvar or supra-valvar PS with indications same as in valvar stenosis, 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 echo is required at each visit; the interval depending on the severity of stenosis.

III. 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 (TOF)

Diagnostic Work-up: Clinical assessment, pulse oximetry, ECG, X-ray chest, echocardiography, lab investigations (Hemoglobin/Packed cell volume, Fluorescence in situ hybridization for 22q11 deletion in some cases). CT Angiography, cardiac catheterization is performed prior to surgery in select cases.

Medical management *(Class I)*: Maintain Hb >14 g/dL (by oral iron or blood transfusion). Beta blockers to be given in highest tolerated doses (usual dose 1-4 mg/kg/day in 2 to 3 divided doses). Prostaglandin infusion for neonates with significant cyanosis.

Management of cyanotic spell: Oxygen administration, knee-chest position, intravenous fluid bolus of normal saline at the rate of 10-20 mL/kg, Morphine (0.1-0.2 mg/kg IV), IV Metoprolol (0.1 mg/kg over 5 minutes, can be repeated every 5 minutes provided no hypotension or bradycardia) or short acting Esmolol infusion (50-200 mg/kg/min), sodium bicarbonate 1-2 mEq/kg given IV, blood transfusion if required. For refractory spells, Phenylephrine infusion (2-5 µg/kg/min), IV Ketamine (0.25-1.0 mg/kg bolus dose), general anaesthesia may be needed. Severe refractory cyanotic spell is an indication for emergency surgery/intervention.

Timing of Surgery

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

II. 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 centre's experience *(Class I).*

III. 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)*.

IV. Patients with anomalous left anterior descending artery from right coronary artery crossing the right ventricular outflow tract, who are likely to need right ventricle to pulmonary artery conduit *(Class I)*:

(*a*) <10 kg weight with significant cyanosis: Aorto-pulmonary shunt

(*b*) >10 kg weight: Total repair using conduit, or double barrel approach after two years of age, when the child weighs >10 kg.

Recommendations for Follow-up

I. Asymptomatic patients with no residual lesion but with free pulmonary regurgitation, not requiring intervention, should be followed up 1-2 yearly, life long.

II. Clinical assessment, ECG and echocardiogram is to be done at each visit. Holter monitoring is indicated in patients suspected to have arrhythmia.

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

IV. Cardiac MRI 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.

V. Infective endocarditis prophylaxis is indicated in non-corrected 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.

Ventricular Septal Defect with Pulmonary Atresia (*VSD-PA*)

Anatomical Types

Type A- Short segment valvar 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. Non-confluent branch pulmonary arteries with MAPCA dependent pulmonary blood flow.

Diagnostic Work-up: Clinical assessment, pulse oximetry, ECG, X-ray chest, echocardiography. Additional imaging in the form of cardiac catheterization, CT angiography/cardiac MRI or a combination of these is essential for planning definitive repair. Lab investigations (Hemoglobin/Packed cell volume, Fluorescence in situ hybridization for 22q11 deletion) in some cases.

Medical management same as outlined in section on Tetralogy of Fallot.

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. Presentation with significant cyanosis at <1 year of age: Aorto-pulmonary 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 right ventricle (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: Aorto-pulmonary 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 to PA conduit at 3-4 years.

(*b*) Suboptimal pulmonary blood flow with small PAs – Additional shunt followed by total repair with RV to PA conduit at 3-4 years.

(*c*) Increased pulmonary blood flow with large PAs – Total repair with RV to PA conduit by 1 year.

Type C (Long segment pulmonary atresia with confluent branch pulmonary arteries supplied by MAPCAs) *(Class I)*:

(*a*) Neonatal presentation – Aorto-pulmonary shunt ± Unifocalisation of MAPCAs.

(*b*) After 1st intervention or late presentation: Total

repair with RV to PA conduit and VSD closure at 3-4 years of age.

Type D (Long segment pulmonary atresia with non-confluent branch pulmonary arteries supplied by MAPCAs) *(Class IIa)*: Aorto-pulmonary shunt + Unifocalisation of MAPCAs, followed by total repair with RV to PA conduit and VSD closure at 3-4 years of age.

Recommendations for Follow-up

I. All patients with VSD-PA require life-long follow-up. Clinical assessment, ECG and echocardiogram is required; the interval depending on the nature of repair, residual or additional lesions, symptoms and functional status.

II. Palliated patients need to be seen more frequently if their oxygen saturation is low and to decide for the next intervention.

III. Infective endocarditis prophylaxis is indicated in non-corrected 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 [6].

Transposition of Great Arteries (*TGA*)

Diagnostic Work-up: Clinical assessment, pulse oximetry, ECG, *X*-ray chest, echocardiography, cardiac catheterization (for balloon atrial septostomy or assessment of adequacy of left ventricle for an ASO or to assess pulmonary vascular resistance in late presenters), CT angiography and cardiac MRI (rarely required).

Indications and Timing of Surgery

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

Pre-surgical stabilization (Class I):

I. Start intravenous infusion of Prostaglandin E1 (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.

II. 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. Indications include:

(*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*) Patient 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: Arterial switch operation (ASO) is the best option *(Class I)*.

Timing of surgery: 7 days to 3 weeks, 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 to 2 months: ASO; extracorporeal membrane oxygenator (ECMO) support may be required in some cases *(Class IIa)*.

(*b*) Presenting between 2 to 6 months: ASO with ECMO support or rapid two stage ASO* or an atrial switch (if rapid two stage or ECMO not feasible) *(Class IIa)*.

(*c*) Presenting between 6 months to 2 years: Atrial switch operation (Senning or Mustard operation) *(Class IIa)*. Rapid two stage ASO* to be considered in select cases after detailed evaluation *(Class IIb)*.

**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 aorto-pulmonary shunt as the first stage. The same can also be achieved in select patients by stent placement in a patent ductus arteriosus (Class IIb). It must be noted that ASO with ECMO support and rapid two stage ASO have higher morbidity and mortality than primary ASO.*

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 one year of age.

IV. TGA with VSD and coarctation of aorta: 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*) Subvalvar pulmonary obstruction with normal or near-normal pulmonary valve and pulmonary annulus: ASO with resection of subvalvar 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 >10kg).

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 cavo-pulmonary connection *(Class IIa)*.

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

Double Outlet Right Ventricle (DORV)

Diagnostic Work-up: Clinical presentation, ECG, X-ray chest, pulse oximetry, echocardiography, cardiac catheterization (in select cases), CT angiography and cardiac MRI (when anatomy unclear).

Indication and Timing of Surgery

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

Timing and type of surgery depends on DORV variant (Class I)

I. DORV with subaortic VSD and pulmonary stenosis (TOF type DORV):

(*a*) Presenting with significant cyanosis at <3-4 months: Aorto-pulmonary 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*) Arterial switch operation (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 same sitting.

IV. DORV with subpulmonary VSD and pulmonary stenosis:

(*a*) If pulmonary obstruction is localized *e.g.* subvalvar fibrous membrane or ridge: ASO with resection of subvalvar stenosis.

(*b*) If pulmonary obstruction is tubular or valvar: 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 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.

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. Follow-up visits should include clinical assessment, ECG and echocardiography.

III. Infective endocarditis prophylaxis 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 (ccTGA)

Diagnostic Work-up: Clinical assessment, pulse oximetry, ECG, *X*-ray chest, echocardiography, cardiac catheterization (in select cases), cardiac MRI (in adults or after surgery), electrophysiological testing (selected patients, who have arrhythmias/blocks).

Indications and Timing of Surgery [7,8]

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 left ventricle is functioning at systemic pressure and when such surgery is feasible *(Class IIa)*.

Indications and Timing for Specific Groups of ccTGA

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 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 (pulmonary stenosis): Double switch (atrial switch plus Rastelli) *(Class I)* or univentricular repair pathway *(Class IIa)*. If the saturation is good, medical follow-up may be considered after discussion with the family.

IV. Associated with complete heart block: Permanent, dual chamber pacemaker implantation *(Class I)*.

Recommendations for Follow-up

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

II. Infective endocarditis 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*)

Diagnostic Work-up: Clinical assessment, ECG, *X*-ray chest, pulse oximetry, echocardiography, cardiac catheterisation, CT angiography, cardiac MRI.

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 [9]. Treating physician must inform and discuss the details with the parent/ guardian prior to 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:

(*i*) Type of surgery: Pulmonary artery banding at 4-6 weeks of age, preferably before 3 months.

(*ii*) Additional procedures may be required if systemic outflow obstruction is present.

(*b*) With decreased pulmonary blood flow (pulmonary stenosis group): Systemic to pulmonary artery shunt or stenting of ductus arteriosus if systemic arterial saturation is consistently below 70%-75%.

(*c*) With balanced pulmonary circulation: The baby usually maintains saturations above 80% and is not in failure. Such infants should be followed up closely. Surgery if saturation falls below 70%. *(Class I)*.

II. Those presenting later in life or have undergone first surgery earlier:

(*a*) With pulmonary hypertension and no pulmonary stenosis: 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 pulmonary vascular resistance.

(*b*) With normal pulmonary pressure and resistance due to pulmonary stenosis/previous pulmonary artery banding/previous aorto-pulmonary shunt:

(*i*) Bidirectional Glenn procedure between 4-12 months of age *(Class I)*.

(*ii*) Total cavo-pulmonary connection or completion of Fontan procedure (preferably extracardiac): Between 4-7 years of age when the child weighs 15-20 kg. Fenestration of Fontan circuit is indicated in high-risk cases.

Recommendations for Follow-up

I. All patients with univentricular heart (operated or unoperated) require lifelong follow-up. Frequency should be individualised, but should be at least once a year in stable cases.

II. 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 [9].

III. The threshold for performing cardiac catheterisation during follow-up should be low as a number of complications can be successfully treated if diagnosed in time.

IV. Infective endocarditis prophylaxis 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.

Persistent Truncus Arteriosus

Classification of truncus arteriosus (Van Praagh and Van Praagh's) [10]

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 patent ductus arteriosus or collaterals from the aorta.

IV. Type A4 - There is associated obstructive lesion of the aortic arch.

Diagnostic Work-up: Clinical assessment, pulse oximetry, *X*-ray chest, ECG, echocardiography, CT angiography/cardiac MRI (select cases), cardiac catheterisation (when operability is in doubt).

Ideal Age for Surgery: Surgery indicated in all, unless 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)*.

Type of surgery

Total repair using right ventricle to pulmonary artery conduit. The prospects of repeat surgeries in future for conduit obstruction should be discussed with parents. Truncal valve is repaired if it is regurgitant.

Contraindication for Surgery

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

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.

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

Total Anomalous Pulmonary Venous Connection (TAPVC)

Types of TAPVC

Type I: Anomalous connection at supracardiac level (to innominate vein or right superior vena cava)

Type II: Anomalous connection at cardiac level (to coronary sinus or right atrium)

Type III: Anomalous connection at infradiaphragmatic level (to portal vein or inferior vena cava)

Type IV: Anomalous connection at two or more of the above levels.

Diagnostic Work-up: Clinical assessment, pulse oximetry, ECG, *X*-ray chest, echocardiography, cardiac catheterization (Rarely performed when operability is in doubt), CT angiography/cardiac MRI (select cases).

Indications and Timing of Surgery (all are *Class I* recommendations)

I. Patients with obstructive TAPVC should undergo emergency surgery.

II. Surgery should be performed as early as possible in non-obstructive TAPVC, even if they are asymptomatic.

III. Those presenting late should be evaluated for onset of pulmonary vascular disease and operated if the data suggests operable status.

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.

II. Since arrhythmias can occur long after TAPVC surgery, parents/patients should be informed to report if any symptom suggestive of arrhythmia develops.

Infective endocarditis prophylaxis is indicated in non-corrected 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

Diagnostic Work-up: Clinical assessment, pulse oximetry, ECG, *X*-ray chest, echocardiography, cardiac catheterization (in select cases), cardiac MRI (important when planning surgical repair), electrophysiological studies (select cases).

Indications and Timing for Treatment

Presentation in neonatal period: Significant cyanosis: IV Prostaglandin infusion; Heart failure: Anti-failure therapy including diuretics; Tachyarrhythmias: Antiarrhythmic drugs; Surgery for neonates not stabilized with medical therapy *(Class IIa).*

Presentation in older children and adults: Surgery is indicated *(Class I)* in those with symptoms or deteriorating exercise capacity, cyanosis (oxygen saturation <90%), paradoxical embolism, progressive cardiomegaly on chest *X*-ray (cardiothoracic ratio >0.65), progressive dilation or dysfunction of the right ventricle on echocardiography.

Types of Surgery: Depends on the underlying anatomy and size of the functional ventricle. Options include tricuspid valve repair (Cone repair, best done at about 2 years of age)/replacement (if repair not possible), and one and a half ventricle repair.

Recommendations for Follow-up

I. ECG, *X*-ray chest and echocardiography should be done at each visit. Holter, exercise testing and cardiac MRI may be required in select patients.

II. Asymptomatic patients who are not candidates for surgery can be followed up every 2-3 years.

III. Infective endocarditis 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 aortic regurgitation (AR) occur most commonly secondary to acute or chronic rheumatic heart disease, and they may co-exist in some. Congenital MR is uncommon, however congenital AR due to a congenitally bicuspid aortic valve is not rare.

Diagnostic Work-up: Clinical assessment, ECG, *X*-ray chest, echocardiography, exercise test (in select cases), CT angiography or cardiac MRI (in select cases).

Medical Therapy

I. Angiotensin converting enzyme inhibitors are indicated in patients with severe MR and severe AR. Diuretics to be used in those with dyspnea due to heart failure.

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

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

IV. 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 [11,12]

I. Symptomatic patients with moderate to severe MR

with left ventricular ejection fraction >30% *(Class I)*.

II. 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 >50mmHg.

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

Aortic regurgitation [11]

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 patient 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 [13]

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 non rheumatic AR (if expertise available).

(*b*) Bioprosthetic valve for: female patients planning pregnancy in future, or if compliance with oral anticoagulation is dubious.

(*c*) Prosthetic metallic valve replacements for the rest of patients.

Anticoagulation after Valve Surgery [14]

I. Oral anticoagulant drug: Warfarin or other anticoumarin drug

(*a*) Desired INR (International Normalized Ratio):

(*i*) After mitral valve replacement: 3.0 (±0.5)

(*ii*) After aortic valve replacement: 2.5 (±0.5)

(*iii*) 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 and the warning signs of overdose of warfarin. 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 with atrial fibrillation.

II. Aspirin: Dose - 3 to 5 mg/kg/day given in addition to anticoagulation *(Class I)*.

(*a*) Duration: Valve repair, bioprosthetic valve: For 6 months after surgery

(*a*) Prosthetic metallic valve: Lifelong

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 at periodic intervals.

III. Operated patients with no residual abnormality: Clinical assessment, ECG and echocardiography. Patients with prosthetic metallic valve require frequent monitoring of INR and fluoroscopy (for valve motion).

Infective endocarditis prophylaxis [14]: 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 or periapical region of teeth, or perforation of the oral mucosa, in patients with prosthetic valve and also in those where prosthetic material is used for valve repair (*e.g.* annuloplasty rings).

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