5.8 Congenital Heart Disease

Congenital heart diseases (CHD) refer to structural or functional heart diseases, which are present at birth. Some of these lesions may be discovered later.

Prevalence of CHD

The reported incidence of congenital heart disease is 8-10/1000 live births according to various series from different parts of the world. It is believed that this incidence has not changed much over the years. Nearly 33% to 50% of these defects are critical, requiring intervention in the first year of life itself. With a believed incidence rate of 6-8 per 1000 live births; nearly 180,000 children are born with heart defects each year in India. Of these, nearly 60,000 to 90,000 suffer from critical cardiac lesions requiring early intervention. Approximately 10% of present infant mortality in India may be accounted for by congenital heart diseases alone.

CHD Guidelines

The Working Group on Management of Congenital Heart Diseases in India have devised guidelines for the Management of Congenital Heart Diseases in India published in 2008 following a National Consensus Meeting held on 26th August 2007 at AIIMS.

These guidelines would help determine:

A. Documentation of the pre-operative investigations which are mandatory for authorization prior to surgery.

B. Ideal age for intervention or timing of surgery.

C. Cases where surgery is not indicated.

D. Chances of spontaneous improvement and by which age.

Every pediatrician/ cardiologist/ other health care provider must strive to get a complete diagnosis on a child suspected of having heart disease, even if that requires referral to a higher center.

These guidelines are meant to assist the health care provider for managing cases with congenital heart diseases. While these may be applicable to the majority, each case needs individualized care based on clinical judgment and exceptions may have to be made.

Categorization of Recommendations for surgery

The recommendations are classified into three categories according to their strength of agreement:

Class I: General agreement exists that the treatment is useful and effective.

Class II: Conflicting evidence or divergence of opinion or both about the usefulness/ efficacy of treatment.

IIa: Weight of evidence/ opinion is in favor of heart disease of the primary health care provider usefulness/ efficacy.

IIb: Usefulness/ efficacy is less well established.

Class III: **Evidence and/or general agreement that the treatment is not useful and in some cases may be harmful. These procedures should not be undertaken for surgery as evidence goes against it.**

5.8.1 Atrial Septal Defect (ASD) other than Primum Type

ASD	
Mode of diagnosis: Physical examination, ECG, X-ray Chest, transthoracic echocardiography (transesophageal echo in select cases).	**Spontaneous closure:** Rare if defect >8 mm at birth. Rare after age 2 years. Very rarely an ASD can enlarge on follow up.
Patent foramen ovale: Echocardiographic detection of a small defect in fossa ovalis region with a flap with no evidence of right heart volume over-load (dilatation of right atrium and right ventricle). Patent foramen ovale is a normal finding in newborns.	**Indication for closure:** ASD associated with right ventricular volume overload
Ideal age of closure: *(i)* In asymptomatic child: 2-4 years *(Class I)*. (For sinus venosus defect surgery may be delayed to 4-5 years *(Class IIa)*. *(ii)* Symptomatic ASD in infancy (congestive heart failure, severe pulmonary artery hypertension): seen in about 8%-10% of cases. Rule out associated lesions (e.g., total anomalous pulmonary venous drainage, left ventricular inflow obstruction, aorto-pulmonary window). Early closure is recommended (Class I). *(iii)* If presenting beyond ideal age: Elective closure irrespective of age as long as there is right heart volume overload and pulmonary vascular resistance is in operable range (Class I).	
Method of closure: Surgical: Established mode. Device closure: More recent mode, may be used in children weighing >10 kg and having a central ASD *(Class IIa)*.	

contd.

5.8.2 Atrioventricular Septal Defect (AVSD)

AVSD

Mode of diagnosis:

Physical exam, ECG (left axis deviation of QRS), X-ray chest, echocardiography.

Types:

- Complete form: Primum ASD, Inlet VSD (nonrestrictive), large left to right shunt, pulmonary artery hypertension. Congestive heart failure often present.
- Partial form: Primum ASD with or without restrictive inlet VSD. Congestive heart failure and severe pulmonary hypertension unlikely.

Either type may be associated with variable degree of AV regurgitation or Down's syndrome; early pulmonary hypertension may develop in these children.

Timing of intervention:

- Complete AVSD with uncontrolled congestive heart failure: Surgery as soon as possible; complete repair / pulmonary artery banding according to institution policy (Class I).
- Complete AVSD with controlled heart failure: Complete surgical repair by 3-6 months of age (Class I). Pulmonary artery banding if risk of cardiopulmonary bypass is considered high (Class IIb).
- Partial AVSD, stable: Surgery at about 2-3 years of age (Class I).

Associated significant AV regurgitation may necessitate early surgery.

5.8.3 Ventricular Septal Defect (VSD)

VSD

Mode of diagnosis:

Physical examination, ECG, X-ray chest and echocardiography.

Location of the defect:

Type I: Subarterial (outlet, subpulmonic, supracristal or infundibular)

Type II: Perimembranous (subaortic)

Type III: Inlet

Type IV: Muscular.

Size of the defect:

- ***Large (nonrestrictive):*** Diameter of defect is approximately equal to diameter of the aortic orifice, right ventricular systolic pressure is systemic, and degree of left to right shunt depends on pulmonary vascular resistance.
- ***Moderate (restrictive):*** Diameter of the defect is less than that of the aortic orifice. Right ventricular pressure is half to two third systemic and left to right shunt is >2:1.
- ***Small (restrictive):*** Diameter of the defect is less than one third the size of the orifice. Right ventricular pressure is normal and the left to right shunt is <2:1.

Natural History:

About 10% of large nonrestrictive VSDs die in first year, primarily due to congestive heart failure. Spontaneous closure is uncommon in large VSDs. 30%-40% of moderate or small defects (restrictive) close spontaneously, majority by 3-5 years of age. Decrease in size of VSD is seen in 25%.

VSD

Timing of closure:

(*Class of recommendation:* I, except for the last one)

- Large VSD with uncontrolled congestive heart failure: As soon as possible.
- Large VSD with severe pulmonary artery hypertension: 3-6 months.
- Moderate VSD with pulmonary artery systolic pressure 50%-66% of systemic pressure: Between 1-2 years of age, earlier if one episode of life threatening lower respiratory tract infection or failure to thrive.
- Small sized VSD with normal pulmonary artery pressure, left to right shunt >1.5:1: Closure by 2-4 years.
- Small outlet VSD (<3mm) without aortic valve prolapse: 1-2 yearly follow up to look for development of aortic valve prolapse.
- Small outlet VSD with aortic valve prolapse without aortic regurgitation: Closure by 2-3 years of age irrespective of the size and magnitude of left to right shunt.
- Small outlet VSD with any degree of aortic regurgitation: Surgery whenever aortic regurgitation is detected.
- Small perimembranous VSD with aortic valve prolapse with no or mild aortic regurgitation: 1-2 yearly follow up to look for any increase in aortic regurgitation.
- Small perimembranous VSD with aortic cusp prolapse with more than mild aortic regurgitation: Surgery whenever aortic regurgitation is detected.
- Small VSD with more than one episode of infective endocarditis: Early VSD closure recommended.
- Small VSD with one previous episode of infective endocarditis: Early VSD closure recommended *(Class IIb)*.

Method of closure:

- Surgical closure.
- Device closure for muscular VSD in those weighing >15 Kg. *(Class IIa)*. For peri-membranous VSD *(Class IIb)*.
- Pulmonary artery banding is indicated for multiple (Swiss cheese) *(Class I)*, or very large VSD, almost single ventricle *(Class IIa)*, infants with low weight (<2 Kg) *(Class IIa)*, and those with associated co-morbidity like chest infection *(Class IIb)*.

5.8.4 Patent Ductus Arteriosus (PDA)

PDA

Mode of diagnosis:

Physical examination, ECG, X-ray chest and echocardiography.

table contd.

Size of PDA:

Large PDA: Associated with significant left heart volume overload, congestive heart failure, severe pulmonary arterial hyper-tension. PDA murmur is unlikely to be loud or continuous.

- Moderate PDA: Some degree of left heart overload, mild to moderate pulmonary artery hypertension, no/mild congestive heart failure. Murmur is continuous.
- Small PDA: Minimal or no left heart overload. No pulmonary hypertension or congestive heart failure. Murmur may be continuous or only systolic.
- Silent PDA: No murmur, no pulmonary hypertension. Diagnosed only on echo Doppler.

Spontaneous closure:

Small PDAs in full term baby may close up to 3 mo of age, large PDAs are unlikely to close.

Timing of closure:

- Large/ moderate PDA, with congestive heart failure, pulmonary artery hypertension: Early closure (by 3-6 months) (*Class I*).
- Moderate PDA, no congestive heart failure: 6 months-1 year (Class I). If failure to thrive, closure can be accomplished earlier (*Class IIa*).
- Small PDA: At 12-18 months (*Class I*).
- Silent PDA: Closure not recommended (*Class III*).

Mode of closure:

Can be individualized. Device closure, coils occlusion or surgical ligation in children >6 months of age. Surgical ligation if <6 months of age. Device/ coils in <6 months (*Class IIb*). Indomethacin/ ibuprofen not to be used in term babies (*Class III*).

PDA in a preterm baby:

- Intervene if baby in heart failure (small PDAs may close spontaneously).
- Indomethacin or Ibuprofen (if no contraindication) (*Class I*).
- Surgical ligation if above drugs fail or are contraindicated (*Class I*).
- Prophylactic indomethacin or ibuprofen therapy: Not recommended (*Class III*).

5.8.5 Coarctation of Aorta (COA)

COA

Mode of diagnosis:

Femoral pulse exam (may not be weak in neonates with associated patent ductus arteriosus), blood pressure in upper and lower limbs, X-ray chest, echo. In select cases CT angiography/ magnetic resonance imaging may be required.

Timing of Intervention:

- With left ventricular dysfunction / congestive heart failure or severe upper limb hypertension (for age): Immediate intervention (*Class I*).
- Normal left ventricular function, no congestive heart failure and mild upper limb hypertension: Intervention beyond 3-6 months of age (*Class IIa*).
- No hypertension, no heart failure, normal ventricular function: Intervention at 1-2 of age (*Class IIa*).

Intervention is not indicated if Doppler gradient across coarct segment is <20 mmHg with normal left ventricular function (*Class III*).

COA

Mode of intervention:

- Balloon dilatation or surgery for children >6 mo of age.
- Surgical repair for infants <6 mo of age.
- Balloon dilatation with stent deployment can be considered in children >10 years of age if required (*Class IIB*).
- Elective endovascular stenting of aorta is contraindicated for children <10 years of age (*Class III*).

5.8.6 Aortic Stenosis (AS)

AS

Mode of diagnosis:

Physical examination, ECG, echocardiography.

Timing of intervention: Valvular AS:

- ***For infants and older children:***
  - Left ventricular dysfunction: Immediate intervention by balloon dilatation, irrespective of gradients (*Class I*).
  - Normal left ventricular function: Balloon dilatation if any of these present:

    (i) gradient >80 mmHg peak and 50 mmHg mean by echo-Doppler (*Class I*)

    (ii) ST-T changes ECG with peak gradient of >50 mmHg (*Class I*)

    (iii) symptoms due to AS with peak gradient of >50 mmHg (*Class IIa*). In case of doubt about severity/symptoms, an exercise test may be done for older children (*Class IIb*).
- **For neonates:**

  Balloon dilatation if symptomatic or there is evidence of left ventricular dysfunction/ mild left ventricular hypoplasia (*Class I*), or if doppler gradient (peak) >75 mmHg (*Class IIa*).

Subvalvular AS due to subaortic membrane:

Surgical intervention if any of the following (Class I): Peak gradient >64 mmHg; or aortic regurgitation of more than mild degree.

5.8.7 Valvular Pulmonic Stenosis (PS)

PS

Mode of diagnosis:

Physical examination, ECG, echocardiography.

table contd.

PS

Timing of intervention: Valvular AS:

- Right ventricular dysfunction: Immediate intervention irrespective of gradient (*Class I*).
- Normal right ventricular function: Balloon dilatation if Doppler gradient (peak) >60mmHg (*Class I*).
- In neonates: Balloon dilatation indicated if right ventricle dysfunction/ mild hypoplasia or hypoxia present (*Class I*).

5.8.8 Tetralogy of Fallot (TOF)

TOF

Mode of diagnosis:

Physical exam, ECG, X-ray chest, Echocardiography. In select cases, cardiac catheterization, CT angio and / or Magnetic resonance imaging may be required.

Medical Therapy:

Maintain Hb >14 g/dL (by using 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).

Timing of surgery:

All patients need surgical repair.

- Stable, minimally cyanosed: Total correction at 1-2 years of age or earlier according to the institutional policy (*Class I*).
- Significant cyanosis (SaO < 70%) or history of spells despite therapy
- <3 months: systemic to pulmonary artery shunt (*Class I*).
- >3 months: shunt or correction depending on anatomy and surgical centers' experience (*Class I*).
- VSD with pulmonary atresia, adequate PAs: Repair at 3-4 years, if right ventricle to pulmonary artery conduit required (*Class I*). Systemic to pulmonary artery shunt if symptomatic earlier and repair without conduit is not possible.

5.8.9 TOF like condition where two ventricular repair is possible (Transposition of the great arteries {TGA} with routable VSD)

TOF like condition where two ventricular repair is possible

Timing of surgery:

For stable cases who are mildly blue (*Class I*): repair at 1-2 years of age if conduit not required; repair at 3-4 years of age if conduit required. Perform a systemic to pulmonary shunt if the child presents earlier with significant cyanosis (SaO <70%).

5.8.10 TOF like condition where two ventricular repair not possible (Tricuspid atresia, TGA with non-routable VSD)

TOF like condition where two ventricular repair is possible

Timing of surgery:

- Stable, mildly cyanosed: Direct Fontan operation (total cavopulmonary shunt) at 3-4 years (*Class I*).
- Stable, mildly cyanosed: Glenn (superior vena cava to pulmonary artery shunt) at 1 year, Fontan at 3-4 years (*Class IIa*).
- Significant cyanosis (SaO <70%) <6 mo: Systemic to pulmonary shunt followed by Glenn at 9 mo-1 year and Fontan at 3-4 years (*Class I*).
- Significant cyanosis (SaO <70%) >6 mo: Bidirectional Glenn followed by Fontan at 3-4 years of age (*Class I*).

5.8.11 Transposition of Great Arteries (TGA)

TGA

Mode of diagnosis:

Physical exam, X-ray chest, Echocardiography.

Balloon atrial septostomy:

Indicated (if ASD is restrictive) in: TGA with intact ventricular septum (*Class I*); TGA with VSD and/or PDA if surgery has to be delayed for a few weeks due to some reason (*Class IIa*).

Timing of surgery:

- TGA with Intact interventricular septum
  - If <3-4 wks of age: Arterial switch immediately (Class I).
  - If >3-4 wks of age at presentation: Assess left ventricle by echo. If the left ventricle is decompressed: Senning / Mustard at 3-6 mo (Class IIa), or rapid two stage arterial switch (Class IIb). Approach would depend on institutional practice. If the left ventricle is still prepared, very early arterial switch operation (Class IIa) is indicated. In borderline left ventricle: Senning or Mustard (Class IIa); or arterial switch operation (Class IIb) is indicated. Adequacy of left ventricle for arterial switch operation can be assessed by echo in most cases.
- TGA with ventricular septal defect:Arterial switch operation, by 3 months of age (Class I).

5.8.12 Total Anomalous Pulmonary Venous Connection (TAPVC)

TAPVC

Mode of diagnosis:

Physical exam, X-ray chest, ECG and Echo. Cath / CT angio may be required in select cases.

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.

Each type can be obstructive (obstruction at one of the anatomic sites in the anomalous pulmonary venous channel) or non-obstructive. Type III is almost always obstructive.

Timing of surgery:

- Obstructive type: Emergency surgery (*Class I*).
- Non obstructive type: As soon as possible (beyond neonatal period if baby is clinically stable) (*Class I*).
- Those presenting after 2 years of age: Elective surgery whenever diagnosed, as long as pulmonary vascular resistance is in operable range.

5.8.13 Persistent Truncus Arteriosus (TA)

TA

Mode of diagnosis:

Physical exam, X-ray chest and Echo.

Timing of surgery:

Total repair using right ventricle to pulmonary artery conduit. If congestive heart failure remains uncontrolled despite therapy: as soon as possible (*Class I*). If stable, controlled congestive heart failure: by 6-12 weeks of age (*Class I*). The prospects of repeat surgeries for conduit obstruction should be discussed with parents. Pulmonary artery banding if total repair not possible (*Class IIb*).

(Source: Working Group on Management of Congenital Heart Diseases in India. Consensus on Timing of Intervention for Common Congenital Heart Diseases. Indian Pediatr. 2008 Feb; 45(2): 117-126.)