Consensus on Timing of Intervention for Common Congenital Heart Diseases

WORKING GROUP ON MANAGEMENT OF CONGENITAL HEART DISEASES IN INDIA

ABSTRACT

Justification: *Separate guidelines are needed for determining the optimal timing of intervention in children with congenital heart diseases in India, because of their frequent late presentation, undernutrition and co-existing morbidities.* ***Process:*** *Guidelines emerged following expert deliberations at the National Consensus Meeting on Management of Congenital Heart Diseases in India, held on 26th August 2007 at the All India Institute of Medical Sciences, New Delhi, India, supported by Cardiological Society of India.* ***Objectives:*** *To frame evidence based guidelines for (i) appropriate timing of intervention in congenital heart diseases; (ii) assessment of operability in left to right shunt lesions; and (iii) prophylaxis of infective endocarditis in these children.* ***Recommendations:*** *Evidence based recommendations are provided for timing of intervention in common congenital heart diseases including left to right shunt lesions (atrial septal defect, ventricular septal defect, patent ductus arteriosus and others); obstructive lesions (coarctation of aorta, aortic stenosis, pulmonary stenosis); and cyanotic defects (tetralogy of Fallot, transposition of great arteries, total anomalous pulmonary venous connection, truncus arteriosus). Guidelines are also given for assessment of operability in left to right shunt lesions and for infective endocarditis prophylaxis.*

Key words: *Children, Consensus statement, Congenital heart disease, India, Surgery.*

These guidelines originated from a National Consensus Meeting on "Management of Congenital Heart Diseases in India" held on 26th August 2007 at the All India Institute of Medical Sciences, New Delhi, India.

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I. INTRODUCTION

Congenital heart diseases (CHD) refer to structural or functional heart diseases, which are present at birth. Some of these lesions may be discovered later. The reported incidence of congenital heart disease is 8-10/1000 live births according to various series from different parts of the world(1). It is believed that this incidence has not changed much over the years(2). Nearly 33% to 50% of these defects are critical, requiring intervention in the first year of life itself(3).

PREVALENCE OF CHD IN INDIA

We have no community-based data for incidence of congenital heart disease at birth in India. Since a large number of births in our country take place at home, mostly unsupervised by a qualified doctor, hospital statistics are unlikely to be truly representative. The incidence of congenital heart disease varies from as low as 2.25 to 5.2/1000 live births in different studies. There are a few studies of prevalence of congenital heart defects in school children; these are mainly offshoots of prevalence studies for rheumatic fever and rheumatic heart disease(4-9). Since a large number of such defects are critical, leading to death in early life itself, the studies on school children have limited value. Going by the crude birth rate of 27.2/1000 (2001 Census data)(10), the total live births are estimated at nearly 28 million per year. 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. In this way, every year a large no of children are added to the total pool of cases with congenital heart disease. We also have a large number of adult patients with uncorrected congenital cardiac defects, primarily because of lack of health awareness and inadequate health care facilities.

Rapid advances have taken place in the diagnosis and treatment of congenital heart defects over the last six decades. There are diagnostic tools available today by which an accurate diagnosis can be made even before birth. With currently available treatment modalities, over 75% of infants born with critical heart diseases can survive beyond the first year of life and many can lead near normal lives thereafter. However, this privilege of early diagnosis and timely management is restricted to children in developed countries only. Unfortunately, majority of children born in developing countries and afflicted with congenital heart disease do not get the necessary care, leading to high morbidity and mortality. Several reasons exist for this state of affairs including inadequate number of cardiologists, cardiac surgeons, specialized cardiac centers etc. However, perhaps the most important reason is the limited understanding and knowledge of congenital heart disease of the primary health care provider (physician, pediatrician, internist *etc*.).

There are two major reasons why we need separate guidelines for our country. Firstly, the results of surgery may be different from center to center and from centres in the western world. Several of our children are underweight for age and have co morbidities like recurrent chest infections, anemia etc., at the time of cardiac surgery. Secondly, a substantial number of cases present late in the course of the disease. This may mandate certain modifications in the treatment protocol necessary for optimizing the outcome. With this background, a National Consensus Meeting was held for expert deliberations and to reach a consensus for evidence based recommendations.

PREAMBLE

1. 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.
2. The proposed guidelines are meant to assist the health care provider for managing cases with congenital heart diseases in their practice. While these may be applicable to the majority, each case needs individualized care based on clinical judgment and exceptions may have to be made.
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: 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 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.

II. AIMS AND OBJECTIVES

1. To outline the optimal timing of intervention in common congenital heart diseases;
2. To formulate evidence based guidelines for infective endocarditis prophylaxis; and
3. To formulate evidence based guidelines for assessment of operability in left to right shunt lesions.

III. GUIDELINES

1. Atrial Septal Defect (ASD), Other than Primum Type

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(11,12). Rare after age 2 years. Very rarely an ASD can enlarge on follow up (11-14).

Patent foramen ovale: Echocardiographic detection of a small defect in fossa ovalis region with a flap with no evidence of right heart volume overload (dilatation of right atrium and right ventricle). 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(15,16) (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*).

2. Ventricular Septal Defect (VSD)

Mode of diagnosis: Physical examination, ECG, *X*-ray chest and echocardiography.

Size of the defect(17)

- *Large (nonrestrictive):* Diameter of the 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 aortic orifice. Right ventricular pressure is normal and the left to right shunt is <2:1.

Location of the defect(18): Type I: Subarterial (outlet, subpulmonic, supracristal or infundibular), Type II: Perimembranous (subaortic), Type III: Inlet, Type IV: Muscular.

Natural History: About 10% of large nonrestrictive VSDs die in first year, primarily due to congestive heart failure(19). 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%.

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*).

Mode 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*).

3. Patent Ductus Arteriosus (PDA)

Mode of diagnosis: Physical examination, ECG, *X*-ray chest and echocardiography.

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(20) (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*).

4. Atrioventricular Septal Defect (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(21).

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

OBSTRUCTIVE LESIONS

5. Coarctation of Aorta (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 years of age (*Class IIa*).

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

Mode of intervention

- Balloon dilatation or surgery for children >6 mo of age(22).
- Surgical repair for infants <6 mo of age.
- Balloon dilatation with stent deployment can be considered in children >10 years of age if required(23) (*Class IIB*).
- Elective endovascular stenting of aorta is contraindicated for children <10 years of age (*Class III*).

6. Aortic Stenosis (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 in 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.

7. Valvular Pulmonic Stenosis (PS)

Mode of diagnosis: Physical examination, ECG, Echocardiography.

Timing of intervention

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

CYANOTIC CONGENITAL HEART DISEASE

8. Tetralogy of Fallot (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(24).

- Stable, minimally cyanosed: Total correction at 1-2 years of age or earlier according to the institutional policy (*Class I*).
- Significant cyanosis (SaO_2< 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.

9. TOF like Conditions where Two Ventricular Repair is Possible (*double outlet right ventricle* (*DORV*), *transposition of great arteries* (*TGA*) *etc. with routable VSD*).

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_2<70%).

10. TOF like Conditions Where Two Ventricular Repair not Possible (*tricuspid atresia, single ventricle, DORV/ TGA with non-routable VSD*).

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_2 <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_2<70%) >6 mo: Bi directional Glenn followed by Fontan at 3-4 years of age (*Class I*).

11. Transposition of Great Arteries (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 operation 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*).

12. Total Anomalous Pulmonary Venous Connection (TAPVC)

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

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.

13. Persistent Truncus Arteriosus

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*).

14. Other Admixture Lesions with Increased Pulmonary Flow Where Two Ventricular Repair not Possible

Elective pulmonary artery banding by 4-8 weeks of life followed by either; Glenn shunt at 6-12 months and Fontan at 4-5 years of age (*Class I*) or Direct Fontan surgery at 3-4 years of age (*Class IIa*).

15. Guidelines for Infective Endocarditis Prophylaxis(25)

1. Every child with congenital heart disease must be advised to maintain good oral hygiene and a regular dental check up.
2. Unrepaired cyanotic heart diseases are high risk conditions for infective endocarditis, therefore prophylaxis is mandatory.
3. Atrial septal defect (secundum type) and valvular pulmonic stenosis are low risk conditions for infective endocarditis and prophylaxis is not recommended.
4. Other acyanotic congenital heart diseases including a bicuspid aortic valve are moderate risk and prophylaxis is recommended.
5. Repaired congenital heart diseases with prosthetic material need prophylaxis for the first six months after the procedure.
6. Device placement by transcatheter route also requires prophylaxis for the first six months.

7. Prophylaxis is recommended for residual defects after a procedure.

16. Guidelines for Assessing Operability in Shunt Lesions (ASD, VSD, PDA, AVSD)

Clinical evaluation

Age: In general children with shunt lesions age <2 years are operable, including those with severe pulmonary arterial hypertension.

Physical exam: Cardiomegaly, congestive heart failure, presence of S3 and a mid-diastolic flow rumble are indicative of large left to right shunt.

Type of defect: Patients with ASD very rarely go into Eisenmenger state. If pulmonary hypertension is present, its etiopathogenesis may be different.

X-ray chest: Cardiomegaly with pulmonary plethora favors operability.

ECG: Presence of left ventricular volume overload pattern may indicate significant left to right shunt in a case with VSD or PDA.

Echocardiography

Dilatation of left atrium and left ventricle suggests significant left to right shunt in post tricuspid shunt lesions (VSD and PDA). Sub-systemic pulmonary artery pressure, as assessed by Doppler would be against Eisenmenger's syndrome with VSD and PDA.

Cardiac catheterization

The calculated parameters by catheterization like pulmonary vascular resistance (PVR), ratio of PVR to SVR (systemic vascular resistance) have a large number of caveats and therefore should not be taken at their face value in isolation.

Vasodilator testing in catheterization lab using 100% oxygen or oxygen with nitric oxide, again has not been shown to be of proven value and does not add significantly to decision making(26,27). It is important to calculate dissolved oxygen when estimating PVR on 100% oxygen.

Despite these errors, patient with a PVRI (PVR indexed to body surface area) of <6 Wood units and PVR/SVR (ratio of pulmonary to systemic vascular resistance) of <0.25 is considered operable. PVRI over 10 Wood units and PVR/SVR ratio of >0.5 is clearly beyond the operable range. Patients with PVRI and PVR/SVR ratio between these values fall in the gray zone and one may have to either resort to oxygen study (with its attendant scope of error) or correlate with clinical data (*e.g.*, age of the patient, heart size on chest *X*-ray etc.). It is best to refer such cases to centres experienced in dealing with these types of cases.

Lung biopsy

The relationship between PVRI and stage of pulmonary vascular disease on lung biopsy is not linear(28). There is significant individual variability. Some of the young, operable children have been shown to have advanced changes on lung biopsy. Hence lung biopsy has minimal role, if any, in assessing operability in shunt lesions.

When in doubt, do not send patient for surgery as the prognosis of an operated patient with markedly raised PVRI is much worse than an average Eisenmenger syndrome patient(29-30).

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ANNEXURE: List of Participants with Affiliations

Cardiologists

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Cardiac surgeons

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