

Congenital and Structural Heart Disease: Fixing up of Holes, Wrong Routes and Tracts

Begum NNF¹, Pasha AKMMK²

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Congenital heart disease (CHD) refers to structural or functional heart disease present at birth, even if first discovered later. The incidence of congenital heart disease is the rate that refers to the number of children born with congenital heart disease related to the total number of births over a period of one year. Incidence varies from 6-8/1000 live births in various studies conducted in many centers. In Bangladesh incidence was found up to 25/1000 live birth in one year in a study conducted in CMH Dhaka. Infant mortality rate of 25.6/1000 live births in the country is also contributed a lot by the death of infants from congenital heart disease¹.

There are eight common lesions which account for about 80% of all congenital lesions. They are in descending order of prevalence are ventricular septal defect (VSD) 30%, patent ductus arteriosus (PDA) 10%, atrial septal defect (ASD) 8%, pulmonary stenosis (PS) 8%, coarctation of aorta (CoA) 5-6%, aortic stenosis (AS) 5-6%, transposition of great arteries (TGA) 4-5%. The remaining 20% or so is made of a variety of more rare or complex lesions. The important pediatric cardiac milestones are successful PDA ligation in 1938 in Boston USA, Coarctation repair in 1945 in USA, ASD closure in 1953 by Gibbon in USA. The life saving intervention of Balloon Atrial Septostomy was first performed in late 1960 by Dr. Rashkind in Philadelphia. In late 1970 introduction of prostaglandin for treatment of ductus dependent pulmonary and systemic circulation provides a measure of securing adequate oxygenation in a number of blue neonate, thus delaying emergency intervention and surgery by keeping them alive. On the other hand indomethacin is used to close the hemodynamically significant PDA in new born. First PDA coil occlusion was performed in 1967 and first ASD device closure was done by King and Mill in 1976. First balloon valvuloplasty was introduced in 1979. First VSD device closure was performed in 1990s. In Bangladesh, all kind of first ever interventions were performed in pediatric cardiology department of CMH Dhaka since 1998^{2,3}.

Our Perspective:

As incidence of CHD is increasing, the magnitude of the problem is becoming enormous in the country. The existing number of trained personnel for pediatric cardiology and pediatric cardiac surgery is inadequate. There was not a single trained person in the country before, a team composed of pediatric cardiologist, cardiac

surgeon, cardiac anesthesiologist, group of cardiac technicians started work in CMH Dhaka after training from Prince Sultan Cardiac Centre, Riyadh, KSA in August 1998 after training of two years. CMH Dhaka is the only center in Bangladesh where cardiac interventions for congenital heart lesions in any age group along with adult congenital heart diseases are available since then. Various patients who need transcatheter interventions are referred to pediatric cardiologist for help from many center of the country. As these treatment is not available in other centers, people has to travel abroad and spend foreign currency to get treatment. Considering the fact civilian non-entitled patient are eligible to get this treatment in CMH Dhaka after permission from the authority^{1,3}.

Life saving treatment for patient of congenital heart disease in Pediatric Cardiology Unit:

Our aim is to save every single life who come to us for help and we never count office hour or lack of manpower support when need to fight with emergency. Examples of emergency interventions are:

- PDA closure in preterm newborn by injection Indomethacin, Ibuprofen, Acetaminophen.
- Prostaglandin infusion for PDA dependent complex cyanotic heart diseases.
- Balloon Atrial Septostomy to maintain cardiac output or to provide mixing.
- Balloon Valvoplasty for critical pulmonary and aortic stenosis.
- Balloon Angioplasty for critical coarctation of aorta.
- PDA stenting in case of pulmonary atresia, extreme Tetralogy of Fallot etc⁴.
- Perforation of pulmonary valve with laser or wire in case of pulmonary atresia.

Transcatheter Intervention in children (Fixing up of holes, wrong routes and tracts)

This is the most challenging part of pediatric cardiac care (curing patient without cutting chest).

- The range of interventional treatment options for structural heart diseases including congenital & valvular heart diseases has developed rapidly over the last few years.

1. **Brig Gen Nurun Nahar Fatema Begum**, SBP, FCPS, FRCP, FACC, FSCAI, Professor and Head, Department of Paediatrics, AFMC, Dhaka & Advisor Specialist in Paediatric Cardiology, CMH, Dhaka (E-mail: colfatema@hotmail.com) 2. **Maj Gen AKM Mustafa Kamal Pasha**, SPP, ndc, MPhil, MPH, Commandant, AFMC, Dhaka.

- Catheter based procedures for CHD has emerged as a valuable alternative to medical & surgical therapy in our country since 1998 when a balloon atrial septostomy was performed on a TGA case in our cathlab to save the life of a newborn in early 1999.
- This treatment is replacing the option of cardiac surgery in many cases and threatening to do so for others as parents and patient do not want any scar mark in the chest wall⁵⁻¹⁰.

Following are the lists of diseases which are possible to be cured by transcatheter interventions:

Congenital Heart Interventions

- ASD secundum device closure.
- Sinus venosus ASD stenting
- Congenital VSD device closure
- Pulmonary & coronary fistula device closure.
- PDA device or coil occlusion
- Balloon angioplasty or stenting of coarctation

Structural Heart Interventions in Adults

- Patent Foramen Ovale device closure
- Post MI VSD device closure
- Alcohol Septal Ablation of Hypertrophic obstructive cardiomyopathy (HOCM)
- Left Atrial Appendage Closure with Watchman, ACP or Amulet devices.

Valvular Heart Interventions in children and adults

- Percutaneous Aortic and pulmonary valve Implantation with Melody, Edward sapien, Core valve, Harmony, Altera etc
- Valvuloplasty of aortic pulmonary and mitral valve
- Paravalvar leak closure
- Percutaneous Mitral valve repair by mitraclip, Pascal clip
- Percutaneous Mitral valve replacement with artificial valves e.g. Tandyme valve, Cardia Q TMVR, Highlife TMVR etc, Intrepid TMVR, Fortis TMVR etc.

Examples of Important Lesions Amenable to intervention:

a. Atrial Septal Defect (ASD) Device Closure¹¹

- Constitute 19% of congenital heart disease.
- Patients are usually asymptomatic and may be detected in adult age, are closed by device or surgically at any age.
- First started in Bangladesh (CMH Dhaka) in 2000.
- Total no of cases: 1598.
- Outcome: Embolisation-2, Mortality-0
- New innovation in our centre: Minimalistic approach of device closure.

b. Ventricular Septal Defect (VSD) Device Closure⁶

- VSD is the most common cardiac defects found in children (30%).

- A VSD is rarely iatrogenic secondary to trauma.
- 0.2% of VSD is due to rupture following MI.
- Device closure started in Bangladesh (CMH DHAKA) in 2004.
- Total no of cases: 620
- Outcome: Embolization –nil, Mortality-nil.
- New innovation of technique: Minimalistic approach

c. PDA (Patent Ductus Arteriosus) Device Closure⁵

- Represents 5-10% of congenital heart disease.
- In infants beyond the age of neonatal period, device and coil occlusion has been advocated since 1980s.
- First started in Bangladesh in 1998 in CMH, Dhaka⁵.
- Total no cases: 2575 so far.
- Outcome: Embolization-4, Mortality-nil
- New innovation: Minimalistic approach

d. Patent Foramen Ovale (PFO) closure

PFO is a channel of varying length with 3 components:

1. Right atrial opening
2. Middle region of overlapping
3. Left atrial opening.

It is normally patent in 40% of general population. Indication of PFO Closure:

- Should be closed before causing any second CVA.
- 65% of PFO patient has migraine.
- Risk of thrombo-embolic manifestation is more for sea divers and flyers.
- Total number of cases performed in our centre: 15, First case was performed in a case of migraine in 2006.

e. Left Atrial Appendage (LAA) Closure: Should be closed for the risk of Stroke and Atrial Fibrillation (AF)

Embolic stroke is thought to be associated with left atrial appendage thrombi. 15% of Atrial fibrillation cases has thrombi in LAA. Patient, who needs lifelong warfarin therapy for non valvular AF and are noncompliant, must have LAA closure with ACP or Watchman Device or Amulet Device. We have experience in one case with ACP device in 2010.

f. Percutaneous Pulmonary Valve Implantation (PPVI) with Melody:

PPVI is usually performed in children operated earlier for diseases involving right ventricular outflow tract (RVOT) e.g. Tetralogy of Fallot (TOF), pulmonary atresia, transposition of great arteries with pulmonary stenosis, Truncus arteriosus, Patient with Ross procedure etc. We have started doing pulmonary valve replacement in our country in CMH Dhaka as first ever in South Asia in 2012 and total number of cases done so far is eight. Number is less as valves are very expensive and number of cases required RVOT conduit is Minimum.

Melody or Edward Sapien valve are used in cases where conduit was placed in RVOT during surgery, but most of the TOF cases are repaired with RVOT or transannular patch and self-expandable valves of larger size are required for these kind of cases. Examples are Venous P-valve, Harmony valve, Beizing Med Zenith, Pulsta, Altera etc. Our plan is to start PPVI on native or patched RVOT in future.

Conclusion

Though pediatric cardiac services are still in its infancy in Bangladesh, extensive efforts are taken from many institutions to include pediatric cardiac care program effectively. Fellowship and MD courses are already initiated and hopefully adequate number of specialist will be available to take care of the children suffering from congenital heart disease.

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