Healthy Heart

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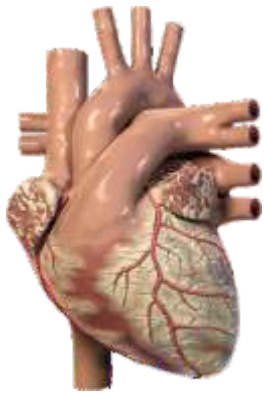

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From the desk of editor:

The field of pediatric cardiology has expanded exponentially in the last few decades. The outlook of children born with structural heart defects has changed dramatically with improved diagnostic facilities, better catheter based or surgical interventional techniques and post operative ICU care. The focus of professionals involved in pediatric cardiology is to provide early definitive care to improve long term outcome in these children. Preventive cardiology in form of fetal diagnosis and molecular genetics will definitely help in curbing the burden of congenital heart disease (CHD) to our society.

With more than 300 successful interventional procedures on children with heart diseases since inception, our department at CIMS has leaped to provide 'state of art care' to children suffering from heart problems. I will be sharing a brief overview of current practice of interventions (surgical or catheter based) in management of common acyanotic CHD with focus on outcome.

- Dr. Kashyap Sheth
Pediatric Cardiologist

Current Trends in Pediatric Cardiology: Catheter and Surgical Interventions in Acyanotic Congenital Heart Diseases (CHD)

Acyanotic CHD are generally divided into two broad groups (I) Left to right shunt lesions and (II) Obstructive lesions.

Left to right shunt lesion (most common group)

Atrial septal defects (ASD) or Partial anomalous pulmonary venous drainage :

Salient Features

1) Presents with right ventricular (RV) volume overload and usually well tolerated in childhood.
2) Most patients present with complications in adulthood – RV dysfunction, pulmonary arterial hypertension (PAH) and atrial arrhythmias beyond 2nd or 3rd decade.
3) Ideal age for closure of ASD is about 3-5 years (pre school age).
4) Some infants (<10%) may present with Congestive Heart Failure (CHF) or severe PAH associated with ASD and demand early closure.

Mode of Closure

1) Device closure: Recommended in centrally located Secundum ASD with good margins.

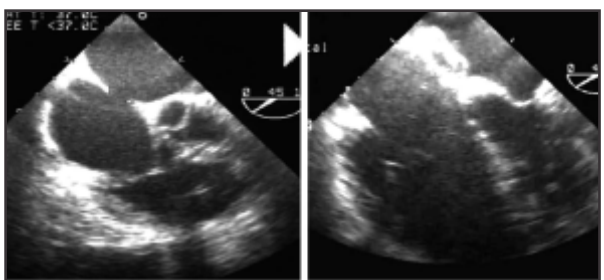

Figure 1. Device Closure of ASD in 6 years old girl with secundum ASD

2) Surgical closure: Recommended in large Secundum ASD with deficient margins, sinus venous or Primum type ASDs.

Ventricular septal defects (VSD):

- Most common CHD amongst all age group.
- Timing and type of intervention in VSD depend on size of defect, location and presence associated malformations & complications.

Large VSD: (VSD size more than 75% of aortic orifice)

1) Associated with CHF, poor growth and severe PAH in infancy. Infants with large VSD usually suffer from frequent Lower Respiratory Tract Infections (LRTI).
2) The risk of significant pulmonary vascular damage increases beyond 6 months of life if large VSD persists. *About 10-20% of such infants die in 1st year of life due to CHF or life threatening LRTI*.
3) Ideal age of intervention in large VSD - within 1 year of life (preferably by 3-6 months). Early repair in infancy carries less risk of permanent pulmonary vascular damage and hence gives good long term outcome.
4) Uncontrolled heart failure or growth failure despite adequate medical management; surgery should be planned earlier.

Moderate VSD: (VSD size 50-75% of aortic orifice)

1) Produces significant volume overload on left heart, PAH is usually insignificant
2) These infants should be followed carefully in the first year of life. If adequate growth is achieved, closure can be planned by 1-2 years of age.

Small VSD: (VSD size less than one third of aortic orifice)

An uncomplicated Small VSD without LV volume overload, no PAH and without Aortic Regurgatation (AR)/ RV Outflow Obstruction (RVOTO) does not need closure. Infective endocarditis prophylaxis should be strictly implemented in these cases.

Indications of closure of small VSD are

1) Occurrence of one episode of infective endocarditis
2) Development of AR due to aortic valve prolapse or RVOTO.

Mode of closure in VSD

1) Surgery: In most type of large VSD, surgery is the preferred mode of intervention. In current era, risk of surgical closure of VSD is less than 5%, much lower than mortality observed in untreated large VSD with CHF in infancy (about 10-20%).
2) Device closure: Percutaneous device closure is generally performed in grown up children (weight >8-10kg) with muscular VSD or post-operative residual defects. Device closure of perimembranous VSD is controversial as it carries risk of heart block (both short term & long term) and aortic valve damage. Hence careful case selection is required to avoid complications.

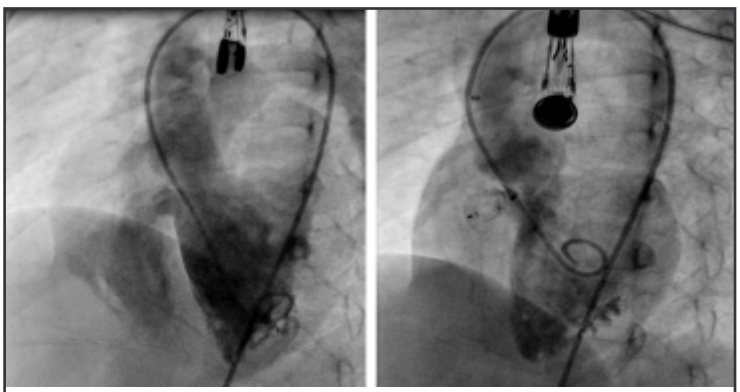

Figure 2. Device closure of VSD through retrograde route in 3 years old girl with perimembranous VSD with significant left to right shunt.

3) Hybrid approach or Perventricular Device Closure: Under certain circumstances, device closure of VSD is performed through puncture in RV free wall.

Situations where this mode is preferred are:

1) Large muscular type VSD associated ventricular dysfunction, to avoid cardiopulmonary bypass
2) Multiple VSDs/Surgically inaccessible VSD where chances of residual defects after surgery are more.

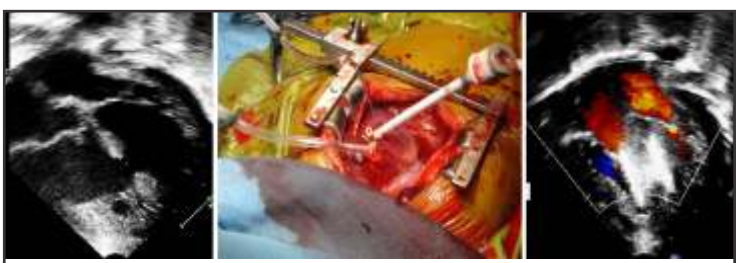

Figure 3. Per Ventricular device closure of VSD in 2 months old infant with large muscular VSD, Severe biventricular dysfunction, severe PAH and CHF. Post procedure hospital stay was 6 days.

Patent Ductus Arteriosus (PDA):

Timing of intervention in PDA largely coincides with VSD; depend on size of PDA and age of patient.

Large PDA:

Usually present with congestive heart failure and poor weight gain in early infancy. Early PDA closure is recommended by 3 to 6 months.

Moderate PDA:

Usually closure indicated by 6 to 12 months of life provided that CHF is controlled clinically and adequate weight gain achieved.

Small PDA:

Closure is recommended after 1 year of age to prevent occurrence of infective endocarditis.

Inaudible PDA:

Insignificant (tiny) PDA which is not audible clinically and documented only on echocardiography does not warrant closure.

Mode of closure in PDA: should be individualized as per size of PDA and patient.

1) Surgically ligation of PDA : Recommended in small infants/ premature babies with large PDA associated with persistent CHF and poor weight gain.
2) Coil or Device closure: Most grown up patients and infants weighing more than 5 to 6 kg can undergo device or coil closure of PDA.

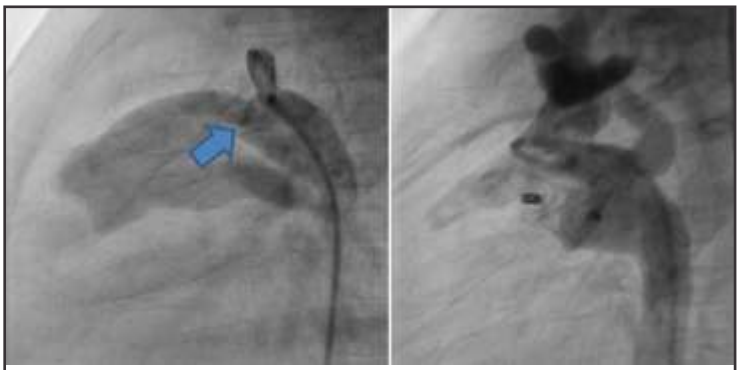

Figure 4. Device closure of large PDA in 4 months old infant weighing 4.5 kg

Other Shunt Lesions:

Aorto-pulmonary window, complete atrioventricular septal defect or ruptured of sinus of valsalva usually produced CHF early in infancy and most of them required surgical repair in first six months of life.

Outcome of Intervention in left to right shunt lesions:

Whether catheter interventional or surgical closure, with proper case selection and timing, *almost all type of left to right shunt lesions carries excellent long term outcome.* With availability of advanced surgical techniques, miniaturization of cardiac interventional hardware and improved post operative care, the chances of recurrent or residual lesions are less and *most patients have survival more than 90-95% on long term.*

Obstructive lesions

Left or right sided obstructive lesions produce pressure overload on ventricles. Significant lesions, if untreated lead to ventricular hypertrophy and dysfunction.

Aortic valve stenosis:

Balloon aortic valvoplasty for severe AS (mean PG > 40mmHg) or moderate AS with symptoms (mean PG > 30mmHg)

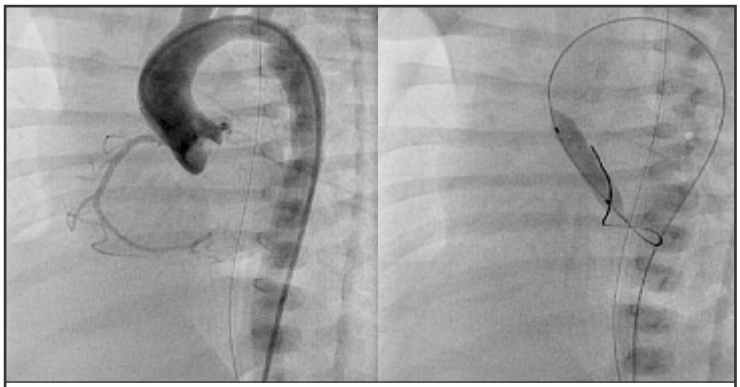

Figure 5. Balloon aortic valvoplasty in 2 days old neonate with critical aortic stenosis with severe biventricular dysfunction and severe PAH.

Pulmonary valve stenosis:

Balloon pulmonary valvoplasty for severe PS (peak PG >60 mmHg) or moderate PS with symptoms (peak PG > 40 mmHg)

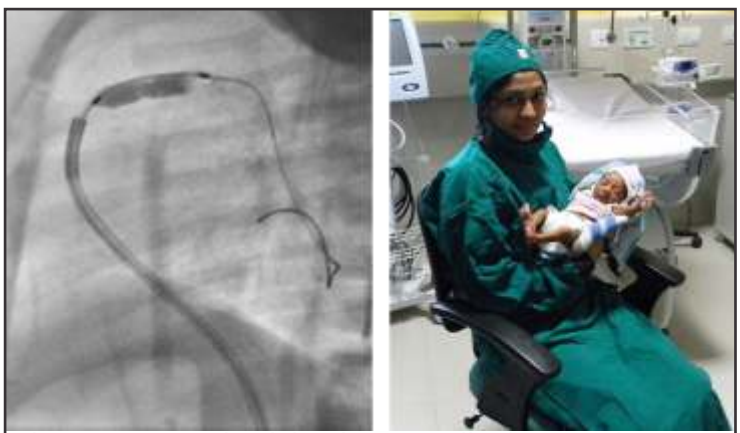

Figure 6. One of the tiniest infant cardiac intervention at our institute. Premature weighing 1.6 kg underwent successful BPV for critical PS. No recurrence till 9 months follow up

Supra or sub valvar aortic or pulmonary stenosis:

Surgery is treatment of choice.

Coarctation of aorta:

Interventions is generally indicated with arch gradient more than 20 mmHg.

Mode of intervention : depends upon multiple indicators.

Balloon dilation: Generally preferred mode of intervention in patients beyond neonatal age group. In critical neonates with severe ventricular dysfunction balloon dilation is preferred over surgery. The balloon dilation of coarctation in neonatal age group carries high incidence of recurrence.

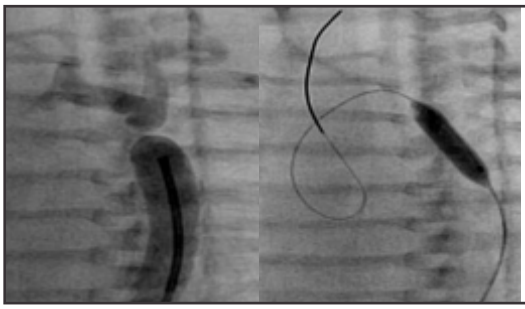

Figure 7. Emergency balloon dilation of coarctation in 7 days old neonate with critical coarctation and severe biventricular dysfunction

Surgery: aortic arch repair is recommended in patients with stable LV function, complex arch anatomy or recurrence following balloon dilation.

Stent placement: Coarctation stenting usually performed in older age group, age more than 8 years, particularly in patients with difficult arch anatomy or recurrent coarctation of aorta.

Outcome in Obstructive Lesions:

Significant obstruction in ventricular outflows generally carries progressive downhill course. Immediate results of surgical repair or balloon valvoplasty in obstructive lesions are excellent. In general, *left side obstructive lesions have tendency for recurrence,* particularly with aortic valve stenosis or coarctation of aorta. So these patients need careful follow up after intervention. *In right sided obstructive problems, the chances of recurrence are less* and patients caries good long term outcome.

Case of the Month

Multidisciplinary Approach in Management of Severe Fetal Cardiac Problem with Excellent Outcome

29 years old lady with 36 weeks gestation was referred for evaluation of fetal cardiac anomaly to our department. Previous fetal cardiac scan done at 34 weeks showed presence of transposition of great arteries with tachycardia and right ventricular enlargement. She had uneventful pregnancy earlier and was on routine supplements. Fetal echocardiography done at our Institute showed normal connection of cardiac chambers and great arteries (AV and VA concordance), restriction of ductus arteriosus (premature ductal closure(PDC)) with severe right heart dilation and dysfunction, severe tricuspid regurgitation, severe tachycardia (FHR = 220-230 /min) and mild pericardial effusion.

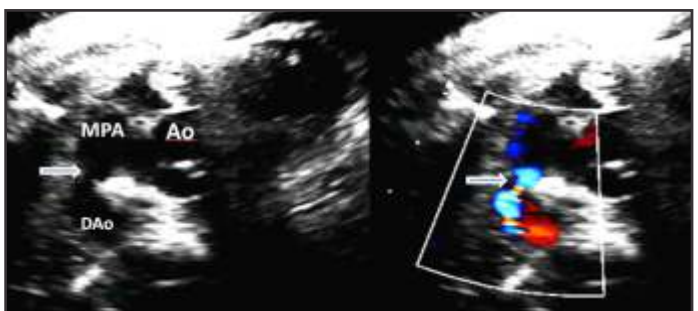

Figure 8. Fetal echocardiography showing short axis view at the level of great vessels. Premature restriction of PDA (arrow) is seen between descending aorta and MPA.

Ductus arteriosus is the only effective outflow from right ventricle in fetal life because the lungs are non functional and receives only 7-10% of total cardiac output in fetal life.PDC leads to severe right heart failure and fetal hydrops if not detected in time. Hence PDC is one of the few cardiac indications of emergency delivery of fetus for survival. An emergency LSCS was performed at our institute within 2 hours of diagnosis of ailment. At birth baby was limp with no respiration and poor tone. She was effectively resuscitated by our NICU team and stabilized. She was transferred to our NICU for further care. Initially she had extreme tachycardia (atrial tachycardia, HR = 220/min) with poor perfusion which was managed with ionotropic support, digitalis and supportive care. She responded well and transferred to ward on 4th day of life. At discharge, she had good right heart function, sinus rhythm and was on breast feeds.

This case signifies the importance of providing correct diagnosis of fetal cardiac malformations and implementation of proper care so that most babies with heart problem will have good neonatal outcome.

Dr. Kashyap Sheth, MD, DNB, FNB
(Pediatric Cardiologist and Interventionist)

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Dr. Kashyap Sheth, MD (PED), DNB, FNB (PED CARDIOLOGY)

Pediatric Cardiologist and Interventionalist; Co-director, Dept. of Pediatric Cardiology, CIMS hospital. Has done Post graduation in Pediatrics from Sheth V.S. Hospital, Ahmedabad and has passed Fellowship Program through National Board of Examination in Ped Cardiology at Escort Heart Institute, New Delhi. He is the first Fellow trained and working exclusively in the field of Pediatric Cardiology in Gujarat state. He has worked as Asst. Professor in Pediatric Cardiology at U.N.Mehta Institute of Cardiology for 3 years and has keen interest and ample of experience in interventional / operative management of sick neonates and infants with congenital heart diseases.

Dr. Amit Chitaliya - MBBS, D.Ped, FPCC (Charite Inst. of Neonatology-Berlin), FPCCC (DHZB-Berlin), FPCCC (NH-India), Fellowship in Pediatric Flexible Bronchoscopy (ERS-France)

Consultant Neonatal & Pediatric intensivist at CIMS Hospital. Graduate and Post graduate(pediatrics)-BJ Medical College, Ahmedabad. Completed "Fellowship in Pediatric Cardiac Critical Care" from Narayana Hridayalaya, Banglore. Has worked as an integral part as senior consultant and developed concept of advanced pediatric critical care at Care Hospital and Lotus Children's Hospital-Hyderabad for over 4 years. He brings along wide experience in neonatology from Charite Institute-Germany and extensive training for HFOV, ECMO and pediatric F.O. Bronchoscopy from German Heart Institute-Berlin & ERS (European Respiratory Society) - France.

Dr. Shaunak B. Shah, MS, MCh, DNB

Pediatric and Adult Cardiac Surgeon. Did his MBBS & MS from Medical College, Baroda. Obtained MCh and DNB in Cardiac Surgery at Sanjay Gandhi Post Graduate Institute of Medical Science, Lucknow. Trained in Congenital Heart Surgery at Chennai under Dr. K. M. Cherian. In addition to Pediatric Cardiac Surgery, his other area of interest is GUCH (Grown up Congenital Heart Disease), Aortic Root problems (Bentall's and Ross procedure) and Post-MI VSDs.

Dr. Milan Chag, MD, DM, DNB

Managing Director, The Heart Care Clinic & CIMS Hospital. Adult, Congenital and Structural Heart Disease intervention specialist. Recipient of National Merit and Post-graduate Merit Scholarships - had extensive training at India's most prestigious institute namely Jaslok Hospital and Research Center, Mumbai, Christian Medical College, Vellore and Sanjay Gandhi Post-graduate Institute, Lucknow. The first cardiologist from Gujarat to possess two highest degrees in cardiology - DM and DNB. One of the most versatile and one of highest volume Interventionalist of India. With 23 years of experience in Cardiology, he has been appointed as a post-graduate teacher in Cardiology by National Board of Examination - the highest medical education body in India. He is the pioneer in establishing pediatric cardiology and pediatric cardiac intervention in Gujarat. He has been a regular faculty at Pediatric Cardiology Society of India. Has been awarded "Lifetime Achievement Award In Cardiovascular Science, Medicine And Surgery" by International Academy of Cardiovascular Sciences, Winnipeg, Canada.

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