Volume - I

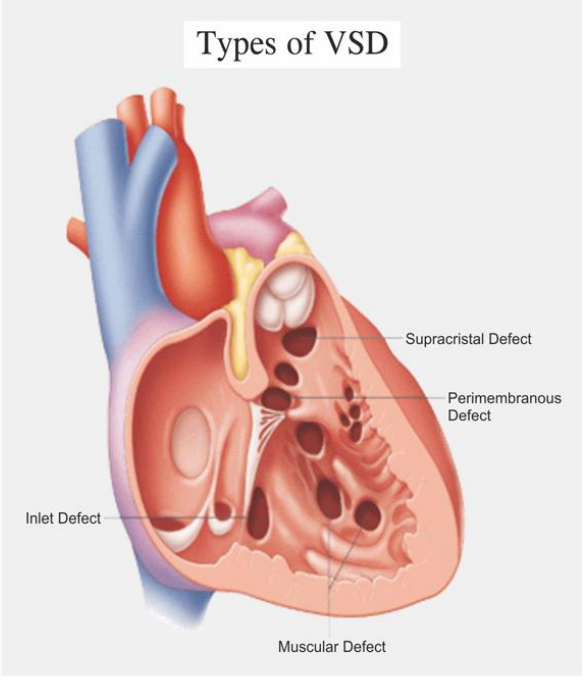
January 2015

The Little Heart News Letter

Advancement in medical therapeutics in pediatric cardiology : Ventricular Septal Defect – a commonest congenital heart disease

Congenital heart disease management is always a challenge worldwide. It is the third commonest cause of death in infancy after prematurity and sepsis. But now with recent advancement in medical technology more than 80% of congenital heart disease can be treated. I Dr Ritesh Sukharamwala am pleased to a inform you that though traditionally surgical correction was considered as gold standard in majority of CHD's now with more research, technology and advancement is treatment various surgical therapeutics are been replaces by interventional non-surgical management.

It is well established that majority of cases of atrial septal defect(ASD) and patent ductus arteriosus (PDA) can be closed without surgery with help of transcatheter DEVICE closure. Ventricular septal defect (VSD) accounts for 25-30% of all CHD's. Now with more understanding of anatomy of VSD and availability of third generation DEVICES majority of moderate size perimembranous VSD are amenable of transcatheter DEVICE closure.

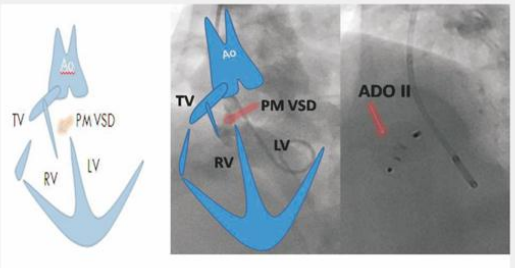


4 Types of VSD : Perimembranous | Muscular | Inlet | Supracristal(subpulmonic)

Case Report 1

A 2 year old boy weighting 7 kg was a known case of moderate perimembranous VSD. Child persists to have poor weight gain with left heart volume overload on cardiac decongestant and adequate diet. This 2D echo showed perimembranous VSD restricted by STL with effective opening of 5.8mm with cardiomeglay(LVIDD-38mm). Child was taken for transcatheter closure of VSD with 6X4 ADO type II device from aortic end without AV looping.

Post procedure child is in sinus rhythm with no residual leak through device At 6 month follow up child is weighing 9.5kg with decrease in left heart size(LVIDD-32mm)



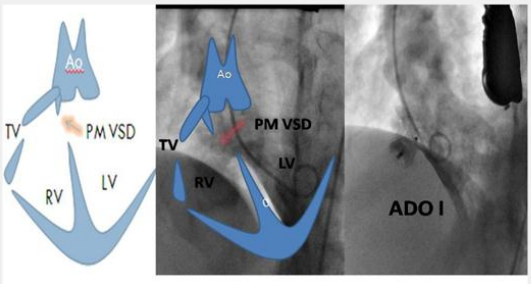
Moderate perimembranous VSD with large septal tricuspid leaflet aneurysm closed with ADO II

Case Report 2

A 1 year old girl weighing 6 kg was diagnosed to have perimembranous VSD type. Despite antifailure measure child was having recurrent episodes of pneumonia and feeding difficulty. 2D ECHO showed large perimembranous VSD measuring 6.5mm with moderate PAH with left heart dilatation(LVID-36mm)

Transcatheter device closure of VSD was done with 8/6 ADO type I device from venous end with AV looping.

Post procedure child is in sinus rhythm with no residual leak through device At 6 month follow up child is weighing 9.0 kg with decrease in left heart size(LVIDD-28mm)



Moderate perimembranous VSD without Septal tricuspid leaflet aneurysm closed with ADO I

Decision Making in Congenital Heart Disease : Redefining OPD Practice

Congenital heart disease (CHD) is a major health problem in India. It accounts for 3rd commonest cause for infant mortality rate (10%). For every 1000 children born 10 children are born with congenital heart disease. In India more than 2 lakhs children are born with congenital heart disease every year and are added to old pool of patients having CHD. Of these around 40-50% of children have critical cardiac problem, requiring intervention in the first year of life itself.

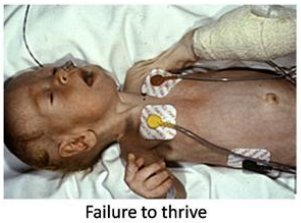
With recently available treatment modalities and timely diagnosis more than 80 % of children born with CHD live near normal life. In developing countries this is not the case due to two reasons namely inadequate number of pediatric cardiologist, cardiac surgeon & specialized centre and poor understanding about CHD among primary health care providers.

When to suspect heart disease in children?

Depending on the severity and type of cardiac lesion child may present with symptoms as early as right from birth or may remain asymptomatic throughout. One should suspect heart disease if child presents with symptoms like repeated lower respiratory tract infection, failure to thrive, feeding disturbances, signs of congestive cardiac failure (CCF)(like tachycardia, tachypnoea, irritability, excessive sweating, hepatomegaly), breathlessness, cyanosis, spells, convulsions or sudden death.

MAJOR : Grade III systolic murmur | Diastolic murmur | Cyanosis | Congestive cardiac failure (CCF)

MINOR : Less than Grade III systolic murmur | Abnormal second heart sound | Abnormal ECG

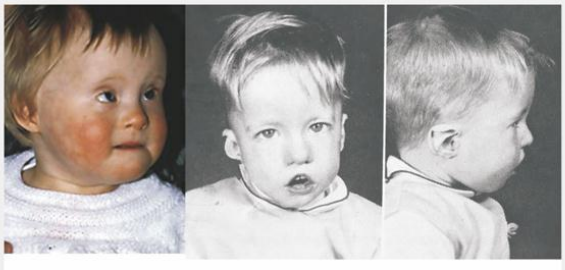


Failure to thrive

In our OPD we can use simple Nada's criteria to qualify any patient for suspecting heart disease. If there is presence of 1 major or 2 minor criteria one should evaluate for heart disease.

If the above said symptoms are present from early childhood or there is documentation of murmur/abnormal chest X-ray/ECG at birth or presence of central cyanosis or extra-cardiac anomalies then the heart disease is likely to be congenital.

If it is congenital heart disease, how to reach differential diagnosis in OPD?



Dysmorphic features

CHD are traditionally classified into Acyanotic or cyanotic CHD. Clinical recognition of cyanotic vs. acyanotic heart disease is made on the basis of physical examination of the patient. The more reliable way of differentiating cyanotic from acyanotic heart disease is by checking the oxygen saturation of the patient with pulse oxymeter. If the child has oxygen saturation of $<94\%$, the child has cyanosis; if the saturation is $> 94\%$ the child has no cyanosis. Incidentally, the human eye cannot recognize cyanosis when the oxygen saturation is between 93 and 85%. Only desaturation below 85% is recognized by the human eye as cyanosis.

Acyanotic and cyanotic congenital heart disease are further sub-classified based on pulmonary blood flow(PBF) into:

- **Normal pulmonary blood flow**
- **Increased pulmonary blood flow**
- **Decreased pulmonary blood flow**

Acyanotic Congenital Heart Disease

- Increased Pulmonary Blood Flow

- Pre-tricuspid shunt (right atrium & ventricle dilated)
  - Atrial septal defect
- Post-tricuspid shunt (left atrium and ventricle dilated)
  - Ventricular septal defect
  - Patent ductus arteriosus
  - Aorto-pulmonary window
  - DORV without PS

- Normal Pulmonary Blood Flow

- Aortic Stenosis
- Pulmonary Stenosis
- Coarctation of aorta

Cyanotic congenital heart disease

- Decreased Pulmonary Blood Flow

- Tetralogy of Fallot
- Pulmonary atresia & variants
- Tricuspid Atresia
- DORV with PS

- Increased Pulmonary Blood Flow

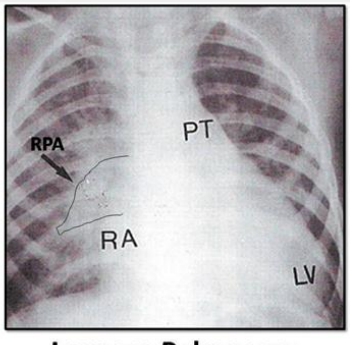
- Transposition of great arteries
- Total anomalous pulmonary venous drainage
- Truncus arteriosus

How to decide on Pulmonary Blood Flow in OPD ?

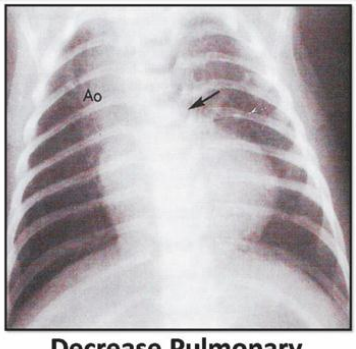
- History
- Chest X-Ray

The **history** of the patient is a key to decide whether the patient has **increased pulmonary blood flow**. The symptoms of feeding difficulty, repeated chest infections, sweating while feeding, baseline tachypnoea, subcostal indrawing and failure to thrive are not symptoms of large left to right shunt but of increased pulmonary blood flow (in both cyanotic and acyanotic heart disease).

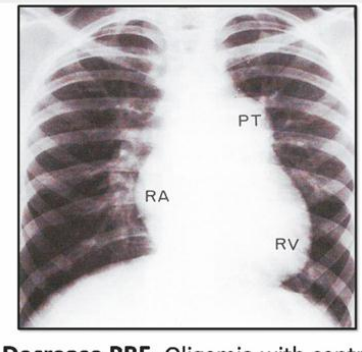
Decreased pulmonary blood flow is on the other hand is evident only when severe, by the presence of severe cyanosis and cyanotic spell. In the older children, squatting, fainting, polycythemia and clubbing are the manifestations of severely decreased pulmonary blood flow.



Increase Pulmonary blood flow - PLETHORA



Decrease Pulmonary blood flow - OLIGEMIA



Decrease PBF- Oligemia with central PA prominent- Eisenmenger syndrome

Deciding on Pulmonary Blood Flow: Chest X-ray :

The chest x-ray is a very specific indicator of the pulmonary blood flow.

Increased PBF

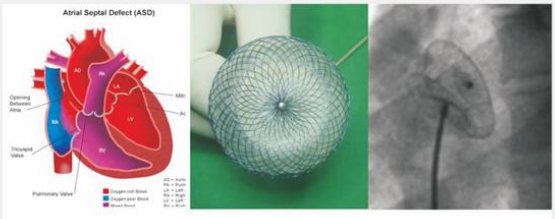
- Dilated and prominent branch pulmonary arteries
- Visible distal branches of pulmonary arteries (outer 2/3rd of lung fields)
- Cardiac Enlargement(cardio-thoracic ratio more than 0.55)
- Fluid in minor fissure
- Congested lung fields

Decreased PBF

- Oligemic lung fields
- Central pulmonary arteries NOT prominent
- When central pulmonary arteries are prominent with oligemic lung fields - Eisenmengers' syndrome.

On the basis of oximeter, it is decided whether the child has cyanotic or acyanotic disease. On the basis of history and chest x-ray, the decision regarding pulmonary blood flow is made. Final evaluation is based on echocardiography. Rarely ECG is needed for confirmation of diagnosis unless arrhythmia is the primary diagnosis. Echocardiography gives a complete anatomical, hemodynamic and functional diagnosis.

How To Manage CHD After Diagnosis? Natural History And Treatment Options



Atrial septal defect(ASD) | Atrial septal occluder device | ASD device closure

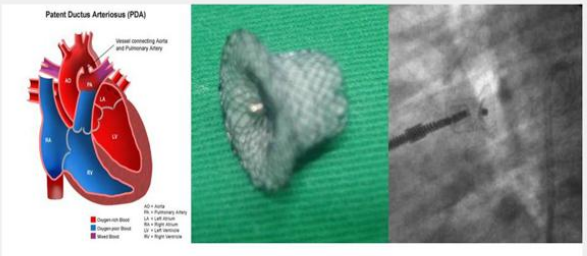
Atrial Septal Defect

ASD is the most benign lesion among the congenital heart disease. Small ostium secundum ASD (< 8mm) usually closes spontaneously by 3-5 years of age. Moderate to large ostium secundum ASD and other types of ASD usually do not close spontaneously. If ASD is treated timely, it gives normal life expectancy. **Ideal age for ASD closure** is 2-4 years of age i.e. preschool age with few exceptions. Most children with an ASD remain active and asymptomatic. Rarely, congestive cardiac failure (CCF) can develop in infancy. If a large defect is untreated, CCF and pulmonary hypertension develop in adults who are in their 20s and 30s. With or without surgery, atrial arrhythmias (flutter or fibrillation) may occur in adults.

With the help of newer techniques more than 80 % of secundum ASD can be closed without surgery i.e. with the help of septal occluder device in same costing as surgery with only one day hospital stay.

Patent ductus arteriosus

PDA is the third most common of congenital heart disease. Any PDA if not closed in 1 month of life never closes spontaneously. Silent PDA [ECHO suggestive of tiny PDA and clinically no murmur] dose not need closure.



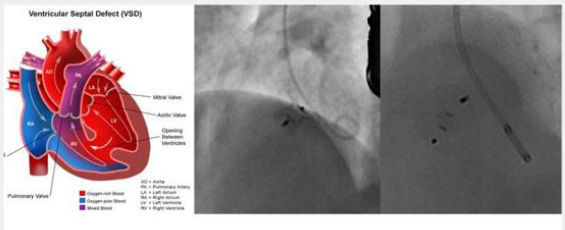
Patent ductus arteriosus (PDA) | Duct occluder device | PDA device closure

Recommended timing for PDA closure

Large PDA needs closure by 3-6 months of age, moderate size PDA needs closure by 6-12 months of age & small PDA should by closed by 1-2 years of age. More than 95 % of PDA can be close safely without surgery with the help of special device called PDA duct occluder.

Ventricular Septal Defect

VSD is the most common congenital heart disease. Large VSD presents as early as 1- 2 month of age with features of CCF, failure to thrive, recurrent respiratory tract infection, excessive sweating, tachypnoea, subcostal indrawing and tachycardia. Small VSD are usually asymptomatic and are diagnosed as incidental finding of murmur.



Ventricular septal defect (VSD) | VSD device closure –ADO I | VSD device closure- ADO II

Natural history of VSD

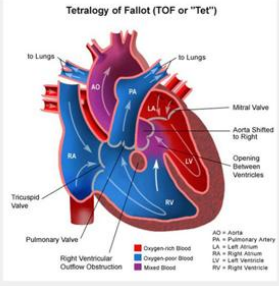
- 99% of small VSD closes spontaneously by 3-5 years of age.
- 85 % of moderate VSD closes spontaneously.
- Large VSD are less likely to become restrictive with time.
- Perimembranous and muscular VSD are more likely to close.
- Inlet VSD, outlet VSD, Malaligned VSD, Swiss cheese VSD never closes.

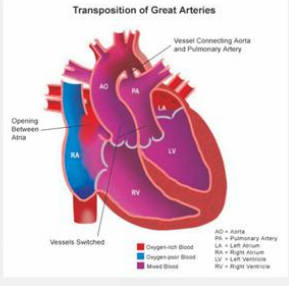
Recommended timing for VSD closure

- Large VSD needs surgical closure by 3-6 month of age depending on symptomatic status of child.
- Moderate VSD if shows feature of failure to thrive and cardiomegaly, needs surgical/ device closure by 1-2 years of age, otherwise natural course of VSD can be followed if child is asymptomatic.
- Asymptomatic small VSD can be followed life long but with quotient for infective endocarditis and newly developing aortic regurgitation (2-4 % of cases).

Tetralogy of Fallot

TOF is the commonest cyanotic heart disease in children. If surgically corrected, they have near normal life expectancy. Corrective surgery for TOF commonly known as intracardiac repair (ICR) is usually done in most centers at 1-2 years of age. If child becomes severely cyanotic and have recurrent episodes of cyanotic spells in early infancy then such patient needs modified BT shunt and ICR can be done later. Those who remain relatively asymptomatic and maintain spO2 > 70-75% in air should undergo ICR directly at 1-2 years of age.



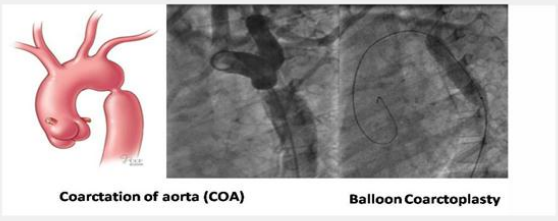


Transposition of Great Arteries (D-TGA)

D-TGA is the commonest cyanotic heart disease occurring in newborn. They manifest with cyanosis from first day of life. Arterial switch operation done timely within 1 month of life, leads to normal life. Timely diagnosis is very much necessary because left ventricle may regress (thinned out) after 4-6 weeks in which case arterial switch operation is not possible.

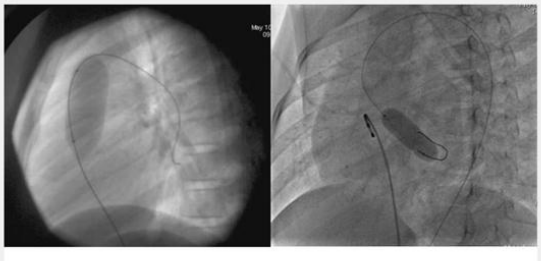
Neonatal Cardiac Emergencies

Symptomatic neonate presenting with features of cyanosis, shock or CCF should be suspected to have CHD after ruling out common neonatal problem. Murmur is usually absent in most of cases. If neonate presents with shock as a predominant feature then it is likely to be left sided obstructive lesions. If neonate presents with cyanosis as a predominant feature then it is likely to be right sided obstructive lesions. If neonate presents with CCF as a predominant feature then it is likely to be mixing lesions



Coarctation of aorta (COA) Balloon Coarctoplasty

- **Left sided obstructive lesions** like Critical AS, Critical COA, Aortic atresia, Hypoplastic left heart.
- **Right sided obstructive lesions** like critical PS, pulmonary atresia intact ventricular septum, pulmonary atresia VSD, Ebstein anomaly
- **Mixing lesions** like d-TGA, Truncus arteriosus, obstructed TAPVC and various complex CHD.



Balloon Pulmonary Valvuloplasty in Pulmonary Stenosis **Balloon Aortic Valvuloplasty in Aortic Stenosis**

Balloon angioplasty in conditions like critical aortic stenosis, critical pulmonary stenosis & critical coarctation of aorta with LV dysfunction are life saving intervention and will lead to near normal life expectancy.

All CHD if diagnosed and intervened timely will have near normal life expectancy with few being high risk. **The myth of 10 kg weight as a criterion for any intervention does not stand anywhere**. Only conditions with single ventricle physiology, complex CHD with multiple defects are likely to have guarded prognosis.

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