

I. ACYANOTIC HEART DISEASES

In acyanotic heart diseases, either there is no abnormal communication between pulmonary and systemic circulation or if such connection is present, the pressure forces the blood from arterial to venous side. The blood supplied to the body is therefore oxygenated & cyanosis does not occur.

A. Disorders of Increased Pulmonary Blood Flow

1. ATRIAL SEPTAL DEFECT

Definition

Atrial septal defects (ASDs) are abnormal openings in the wall separating the right and left atria.

Incidence and Types

Atrial septal defects accounts for about 17% of all congenital cardiac defects and occurs more frequently in girls than in boys.



Atrial septal defects are of 3 types:

a. Ostium Secundum :- This is the commonest type in which abnormal opening is present in the middle of atrial septum.

b. Ostium Primum :- It is also known as partial atrioventricular canal defect. The defect is located in the septum just above the tricuspid valve and is associated with a cleft in the mitral valve and defects of atrioventricular septum.

c. Sinus Venosus :- Sinus venosus ASD is an abnormal opening at the top of atrial septum. The opening may be located adjacent to SVC & is

commonly associated with anomalous connection of the right pulmonary vein to the right atrium or right superior vena cava.

Pathophysiology

- In an ASD, there is communication between the right and left atrium.
- Since, oxygenated blood in left atrium is under high pressure it is forced through the defect into right atrium.
- This left to right shunting of blood places a burden on the right side of heart leading to an increased pulmonary blood flow.
- This shunting of blood does not produce cyanosis; however it leads to pulmonary congestion & right ventricular enlargement.

Clinical Features

The clinical features of ASD are:

- * Children with secundum & sinus venosus atrial septal defects are usually asymptomatic.
- * Rarely there may be congestive heart failure, usually after 3rd or 4th decade of life.
- * The only symptoms may be decreased exercise tolerance and dyspnea.

Diagnostic Evaluation

The diagnosis of ASD is made on the following basis:

- (i) Cardiac Examination - Increased flow of blood across the defect produces a systolic ejection murmur, heard best in the upper left

sternal border. Fixed splitting of second heart sound can be heard in older children. If the left to right shunt is large, a diastolic murmur of increased blood flow across the tricuspid valve may be heard.

(ii) Echocardiogram shows right ventricular volume overload.

(iii) Chest radiograph shows enlargement of the heart & increased pulmonary vascular markings.

(iv) Echocardiogram (2D Echo) can define the location & size of atrial septal defect as well as dilatation of the atria.

Therapeutic Management

Management of atrial septal defect are as follows:

- Small atrial septal defect may occasionally close spontaneously.
- Surgical correction for hemodynamically significant atrial septal defect is recommended between 2-4 years of age. Earlier repair may be done if the child is symptomatic.
- The surgical repair is most often done through a median sternotomy & requires cardiopulmonary bypass.
- If the defect is small, purse string closure is done by stitching around the opening & pulling it closed.
- If the defect is large a knitted Dacron patch is sewn over the defect.

Postoperative Complications

- * Cardiac enlargement

- Date _____
- * Dysrhythmias
 - * Infective endocarditis

2. VENTRICULAR SEPTAL DEFECT

As the children become older left to right shunting of blood no longer occurs. Pulmonary pressure are normal & unoxxygenated blood enters into the systemic circulation.

Definition

Ventricular septal defect (VSD) is an abnormal communication between the right and left ventricle.

Incidence & Types

Ventricular septal defects are the most common heart lesions, accounting for approximately 20% of all congenital heart defects.

Ventricular septal defects are classified according to their size & location in the septum.

a. **Membranous VSDs**: They lie beneath the aortic valve & are most common.

b. **Subpulmonic VSDs**: They lie beneath the pulmonary valve & account for about 5-7% of VSDs.

c. **Atrioventricular Canal Type VSDs or Posterior defects**: They account for approximately 0%.

d. **Muscular VSDs**: They are frequently

Date _____

multiple and represent 5-20% of ventricular septal defects

Pathophysiology

In presence of a ventricular septal defect, a portion of oxygenated blood returning from lungs into the left atrium & left ventricle crosses the ventricular septal defect and enters the right ventricle, from where it returns to the pulmonary circulation. The shunt is left to right.

The magnitude of shunt is determined by the size of ventricular septal defect & amount of pulmonary vascular resistance (PVR) present.

High pulmonary vascular resistance will elevate right ventricular pressure (making it approximate to left ventricular pressure) & decrease shunting across ventricular septal defect.

In the newborn period, pulmonary vascular resistance is still high therefore little shunting may occur at this time & the child may be asymptomatic.

As pulmonary vascular resistance decreases over the first one to two months of life the child may become symptomatic. Due to increased blood in right ventricle, right ventricular hypertrophy occurs.

Clinical Features

The clinical features of VSD depend upon

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the size of defect, the degree of shunting, age of the child & pulmonary vascular resistance.

- With small defects, there is little shunting and infant or child is usually asymptomatic.
- With large VSDs, large left to right shunt occurs & the infant or the child may experience failure to thrive and congestive heart failure.
- Medium sized defects produce symptoms like dyspnoea, tachypnoea, slow physical development, feeding difficulties and frequent pulmonary infections.

Diagnostic Evaluation

The diagnosis of VSD is made on the following basis:

- i) Cardiac examination:** - Blood flows across the ventricular septal defect & produces a systolic murmur that can be heard best at mid to lower left sternal border. In presence of large left to right shunt, the increased regurgitation of blood across mitral valve produces a diastolic low rumble.
- ii) Electrocardiogram:** - In presence of small ventricular septal defect, the ECG is normal. With moderate to large ventricular septal defects, right ventricular hypertrophy is usually present.
- iii) Chest radiograph:** - With moderate to

large size ventricular septal defects, the heart size and pulmonary vascular markings are increased.

Echocardiogram: - Color 2D echocardiography is helpful in determining the size and location of ventricular septal defect. The degree of left to right shunting and PVR can also be assessed.

Therapeutic Management

- 75-80% of small ventricular septal defects & 5-10% of large VSDs will spontaneously close, usually during the first two years of life.
- Infants with small ventricular septal defects require no surgery, except antibiotics to prevent endocarditis.
- Infants with moderate to large ventricular septal defects who are symptomatic, showing signs of congestive heart failure and failing to grow are usually medically managed with a combination of Digoxin and Diuretics. If the infant continues to show signs of congestive heart failure then early surgical repair is indicated.
- Surgical repair of ventricular septal defects is an open heart procedure. VSD of children weighing less than 7 kg are usually repaired with child in deep hypothermia causing circulatory arrest. In older children cardiopulmonary bypass is used. Moderate to small sized

Date _____

VSDs are closed by purse string sutures, while for large defects a synthetic Dacron patch is used to close the defect. The cardiac tissue covers the patch completely within 6 months of surgery. Large ventricular septal defects are closed through a median sternotomy & an incision in the right atrium rather than an incision through the left or right ventricle. Following surgical repair antibiotic prophylaxis for endocarditis is required along with cardiology follow-up.

Complications

Residual ventricular septal defects
Conduction abnormalities.