Ventricular Septal Defect

:Objectives

.Recognise types and pathology of VSD.1

.Identify the clinical manifestations of VSD.2

Describe the physical signs of VSD.3

.Recognise investigations in patients with VSD.4

Explain management of VSD.5

Recognise complications and natral history of.6
.VSD

Prevalence

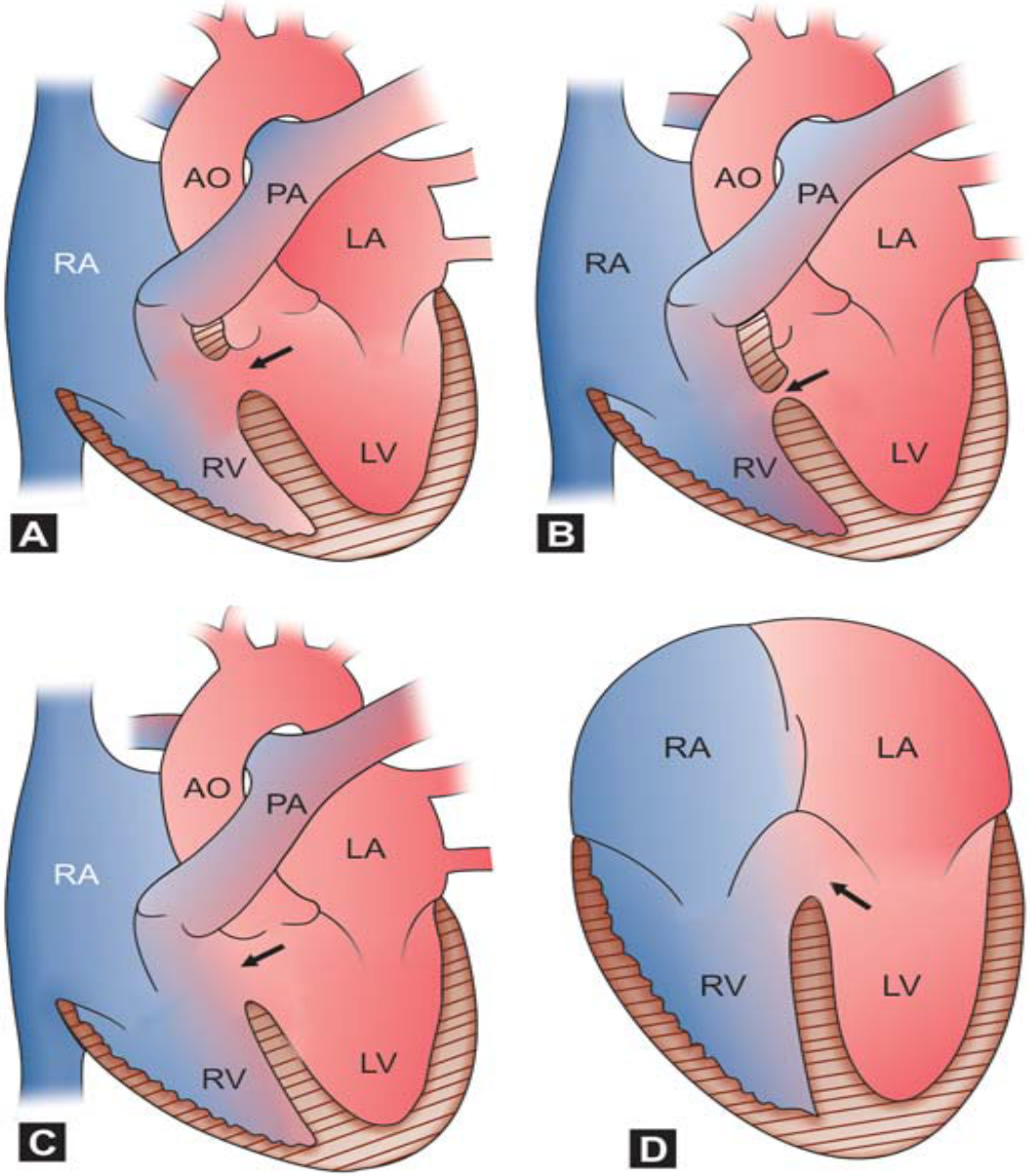
Ventricular septal defect is the most common form of CHD and accounts for 20% to 30% of all such defects, not including those occurring as .part of cyanotic CHDs

DEFINITION

Ventricular septal defect (VSD) is a congenital defect in the interventricular septum, which connects) hole(both ventricles of the heart

Pathology

The ventricular septum may be divided into a small membranous portion and a large muscular portion. The muscular septum has three components: the inlet septum, the trabecular septum, and the outlet (infundibular or conal) septum. The trabecular septum (also simply called the muscular septum) is further divided into anterior, posterior, mid, and apical portions. Therefore, VSD may be classified as a membranous, inlet, outlet (or infundibular), midtrabecular (or midmuscular), anterior trabecular (or anterior muscular, posterior trabecular (or posterior muscular), and apical .muscular defect



Schematic diagrams of different types of VSDs. Arrow showing (A) Sub-aortic ,(Perimembranous) Muscular, (C)) B(Subpulmonic (supracristal), and (D) Inlet type

a. The membranous septum is a relatively small area immediately beneath the aortic valve. The membranous defect involves varying amounts of muscular tissue adjacent to the membranous septum (perimembranous VSD). Perimembranous defects are most common .(70%)

b. Outlet (infundibular or conal) defects account for 5% to 7% of all VSDs in the Western world and about 30% in Far Eastern countries. The defect is located within the outlet (conal) septum, and part of its rim is formed by the aortic and pulmonary annulus. An aortic leaflet can prolapse through the VSD and cause aortic insufficiency (It has been .called a *supracristal, conal, subpulmonary,* or *subarterial defect)*

c. Inlet (or AV canal) defects account for 5% to 8% of all VSDs. The defect is located posterior and inferior to the perimembranous defect beneath the septal leaflet of the tricuspid valve

Muscular: 5%-20%
Central: mid-muscular, may have multiple apparent channels on RV side and coalesce to single defect on LV side
Apical: multiple apparent channels on RV side may be single defect on LV side as with central defect
Marginal: along RV septal junction
Swiss cheese septum: large number of muscular defects

Clinical Manifestations

History

Small VSD

These patients are asymptomatic. The clinical course is benign throughout the infancy even upto old age, but they are at risk for infective endocarditis, which is again rare before age 2 years. Patients with small VSD have normal .growth, normal physical activities and lead a normal life

Moderate to Large VSD

These patients may be symptomatic from birth. Infant is dyspneic irritable but becomes comfortable while held in sitting posture. Dyspnea and diaphoresis occur during feeding indicating presence of congestive heart failure. If untreated some infants may be died due to heart failure and/or pneumonia but most of the infants gradually feel better with medical management and grow with retarded growth. Though symptoms of heart failure improves beyond infancy some children develop dyspnea on effort and recurrent respiratory infection, where as some become almost asymptomatic during childhood. The possible reasons for this asymptomatic group are either VSD has closed or in closing stage or pulmonary vascular disease .has developed restricting pulmonary flow

Large VSD with SeverePulmonary Hypertension

The clinical scenario in this group is quite different from that of large VSD without high PVR. Cyanosis develops from early childhood and subsequently variable degree of cyanosis is present depending on the magnitude of right to .left shunt

First heart sound is loud, second heart sound is closely split and P2 component is very loud (booming character); a diastolic tap is felt (corresponds to loud P2). An ejection click (pulmonary, related to dilatation of pulmonary artery) is present all over the precordium. A short ejection systolic murmur (not pansystolic) or sometimes no systolic murmur is audible. Early diastolic murmur (due to pulmonary regurgitation) over second, third intercostal space in left .sternal border may be present

Physical Examination

Infants with small VSDs are well developed and acyanotic. Before 2. 1
or 3 months of age, infants with large VSDs may have poor weight
.gain or show signs of CHF
A systolic thrill may be present at the lower left sternal border.. 2
.Precordial bulge and hyperactivity are present with a large-shunt VSD
The intensity of the P2 is normal with a small shunt and moderately. 3
increased with a large shunt. The S2 is loud and single in patients with pulmonary hypertension or pulmonary vascular obstructive disease. A grade 2 to 5 of 6 systolic murmur is audible at the lower left sternal
.border .It may be holosystolic(pansystolic murmur) or early systolic

Electrocardiography

.With a small VSD, the ECG findings are normal. 1
With a moderate VSD, left ventricular hypertrophy (LVH). 2 and occasional left atrial hypertrophy (LAH) may be seen, in some cases mild or moderate elevation of right ventricular pressure can result in right ventricular hypertrophy, which is evident in lead V4R or V1 as an rsR pattern with the increasing .in amplitude with increasing right ventricular pressure
With a large defect, the ECG shows biventricular. 3 hypertrophy (BVH) with or without LAH
4. If pulmonary vascular obstructive disease develops, the .ECG shows RVH only

Echocardiography

Two-dimensional and Doppler echocardiographic studies can identify the number, size, and exact location of the defect; estimate pulmonary artery pressure .identify other associated defects; and estimate the magnitude of the .shunt

Radiography

Cardiomegaly of varying degrees is present and involves the left. 1
atrium (LA), left ventricle (LV), and sometimes right ventricle(RV).
.Pulmonary vascular markings increase

The degree of cardiomegaly and the increase in pulmonary vascular markings directly relate to the magnitude of the left-to-right shunt

In pulmonary vascular obstructive disease, the main pulmonary. 2
artery and the hilar PAs enlarge noticeably, but the peripheral lung
.fields are ischemic. The heart size is usually normal

.Natural History

Spontaneous closure of perimembranous and muscular VSDs can. 1 occur. It occurs more frequently with small defects and during the first 6 months of life. About 60% of small to moderate muscular VSDs close spontaneously but not after 8 years of age. About 35% of small perimembranous VSDs close spontaneously but not after 5 years of age. These VSDs do not become bigger with age; rather, they decrease in size. However, inlet defects and outlet (infundibular) defects do not .become smaller or close spontaneously

CHF develops in infants with large VSDs but usually not until 6 to 8. 2 .weeks of age

Pulmonary vascular obstructive disease may begin to develop as. 3 early as 6 to 12 months of age in patients with large VSDs, but the resulting right-toleft shunt usually does not develop until the teenage .years

Infundibular stenosis may develop in some infants with large defects. 4 and result in a decrease in the magnitude of the left-to-right shunt (i.e., .acyanotic TOF), with an occasional occurrence of a right-to-left shunt

COMPLICATIONS

Small VSD

,common complication of small VSD is infective endocarditis
.mainly beyond 2 years of age

Large VSD

,Complications in Large VSD are (i) CHF, (ii) arrhythmias
infective endocarditis, (iv) Eisenmenger's complex,(v)repeated) iii(
chest infection

Management

Medical

Treatment of CHF, if it develops, is indicated with diuretics (with or. 1 without) digoxin for 2 to 4 months to see if growth failure can be improved. Some centers do not use digoxin. Addition of spironolactone may be helpful to minimize potassium loss from diuretics.Concomitant use of an afterload reducing agent, such as captopril, has gained popularity. Angiotensin-converting enzyme inhibitors may raise the serum potassium level, and) ACE(spironolactone or potassium supplementation should be discontinued. Frequent feedings of high-calori formulas, either by .nasogastric tube or oral feeding also can be effective

Anemia if present, should be corrected by oral iron therapy. These measures often allow delay of surgical treatment and may promote spontaneous reduction or closure of the VSD.

No exercise restriction is required in the absence of pulmonary hypertension. 2

Nonsurgical transcatheter closure of selected VSDs is possible in selected patients when the defect is not too close to cardiac valves and when it is difficult to access surgically. 3

Surgical

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İndications for surgical repair in infancy are uncontrolled congestive heart failure, including growth failure or recurrent respiratory infection. Large defects, even in the absence of symptoms, are repaired prior to 2 years of age (often prior to 1 year) if associated with elevated pulmonary artery pressure. Surgery generally is recommended for older, asymptomatic children with normal pulmonary pressure if the pulmonary-to-systemic flow ratio is >2:1. Defects associated with significant left ventricular dilation are often considered appropriate for .repair

There is a lower threshold for operation in patients with VSD and associated aortic insufficiency, especially if the latter is related to aortic cusp prolapse