Congenital Heart Disease (CHD)

先天性心脏病(一)

Department of Pediatrics
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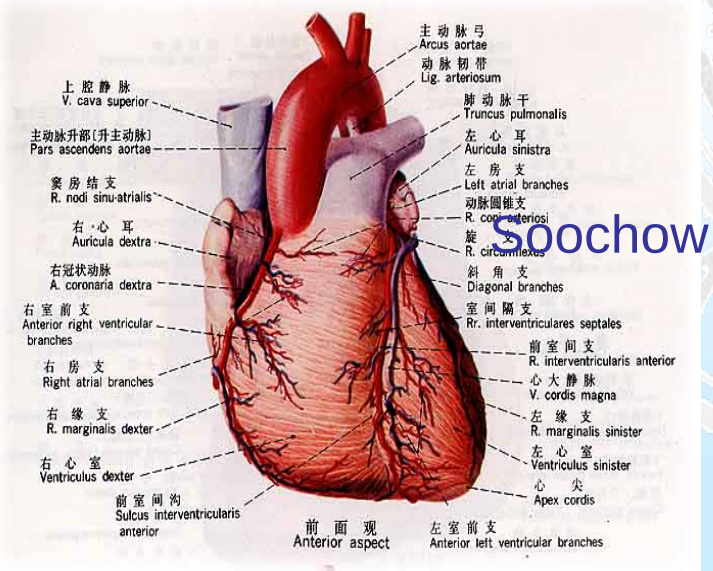


Key words

- congenital heart diseases （CHD）
- lower respiratory tract infection (LRTI)
- Cyanosis
- Ventricular septal defect （VSD）
- Atrial septal defect （ASD）
- patent ductus arteriosus （PDA）
- Pulmonary hypertension （PH）
- Eisenmenger syndrome

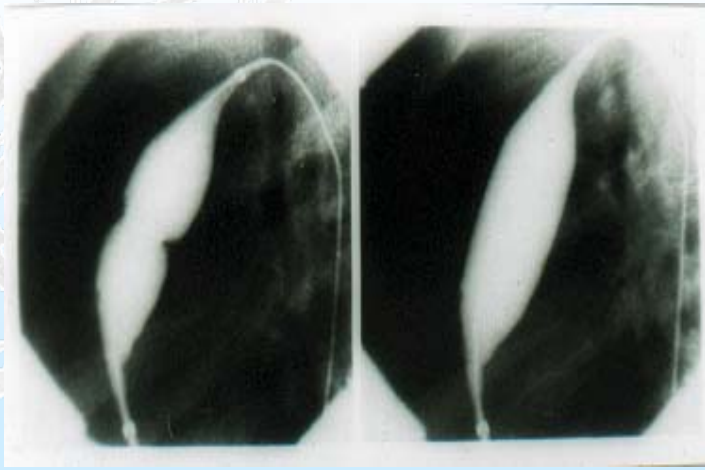
Key words

- squatting
- tetralogy of Fallot （TOF）
- pulmonary stenosis （PS）
- coarctation of aorta （CoA）
- Transposition of great artery (TGA)
- Congestive cardiac failure (CCF)
- Amplatzer Occluder

Overview

1. CHD is defined as an abnormality in circulatory structure malformations present at birth, even if it is discovered much later
2. Major cardiac malformation occur in 6-8/1000 live births and 10-20/1000 have some minor abnormality.

- Diagnosis is aided by the chest X-ray and electricardiogram(ECG),and two-dimension echocardiogram, Great progressive have made in cardiac catheterization,intervention therapy and surgical repair in recent years.



Learning Objective

1. Familiar with the etiology and classification of the common CHD
2. Know the hemodynamic change and the signs, symptoms , diagnostic features and management of the common CHD(VSD,ASD, PDA and TOF)

Etiology

Genetic factor (internal factor)

- CHD result from gene mutation or chromosome aberration

  遗传因素(内在因素) 与基因突变、染色体畸变有关

Environmental factor (external factor)

- CHD is mainly correlation to the intrauterine infection

  环境因素(外界因素) 与宫内病毒感染有关

Prevention

1. **The health protection of pregnant woman should be enhanced**加强孕妇保健
2. **High risk factors should be avoided**避免接触药物、辐射等高危因素
3. **Suit dosage Folic Acid should be filled up in early pregnancy stage**妊娠早期适量补充叶酸

Classification

1. **left-to-right shunts (acyanosis)**

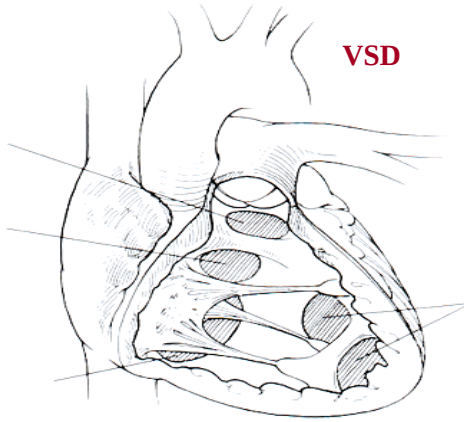
   VSD, ASD, PDA左向右分流型(潜在青紫型)

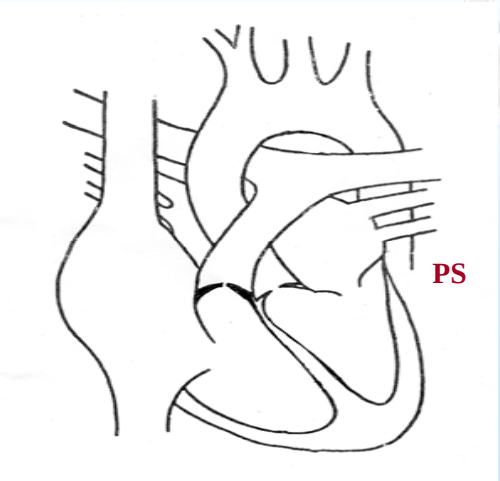
2. **right-to-left shunts (cyanosis)**

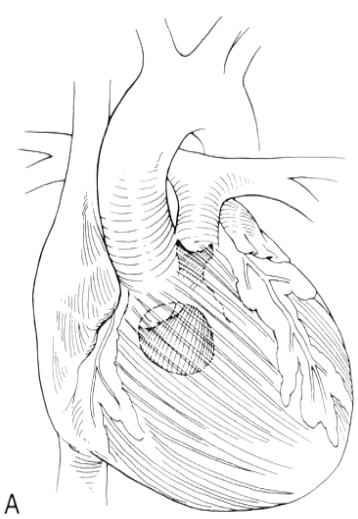
   TOF,D-TGA　右向左分流型(青紫型)

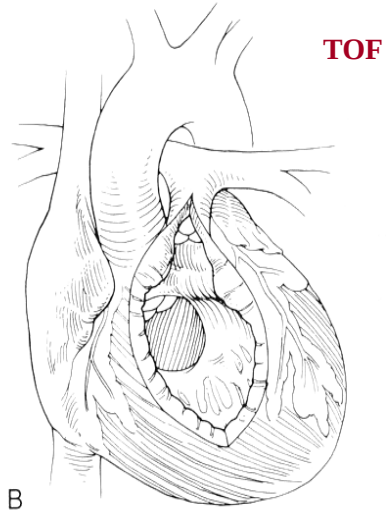
3. **non shunts (noncyanosis)**

   PS , AS, CoA　(无分流型)









Common CHD in Clinic

- **Ventricular septal defect (VSD)**
  室间隔缺损
- **Atrial septal defect (ASD)**
  房间隔缺损
- **Patent ductus arteriosus (PDA)**
  动脉导管未闭
- **Tetralogy of Fallot (TOF)**
  法洛四联症

Clinic Symptoms in CHD

1. Asymptomatic/no symptom 轻症时无症状
2. Difficulty with feeding 喂养困难
3. Recurrent lower respiratory tract infection (LRTI) /chest infection 反复呼吸道感染
4. Dyspenia / shortness of breath /breathlessness on exertion 呼吸困难（气急）
5. Excessive sweating on exertion or with feeds 多汗
6. Failure to thrive / Poor weight gain 生长迟缓
7. Cyanotic spells /Hypoxemic spells/blue spell/paraxysmal hypercyanotic episodes 缺氧发作
8. Squat suddenly (to ward off cyanotic spells) 蹲踞

Atrial septal defect (ASD)

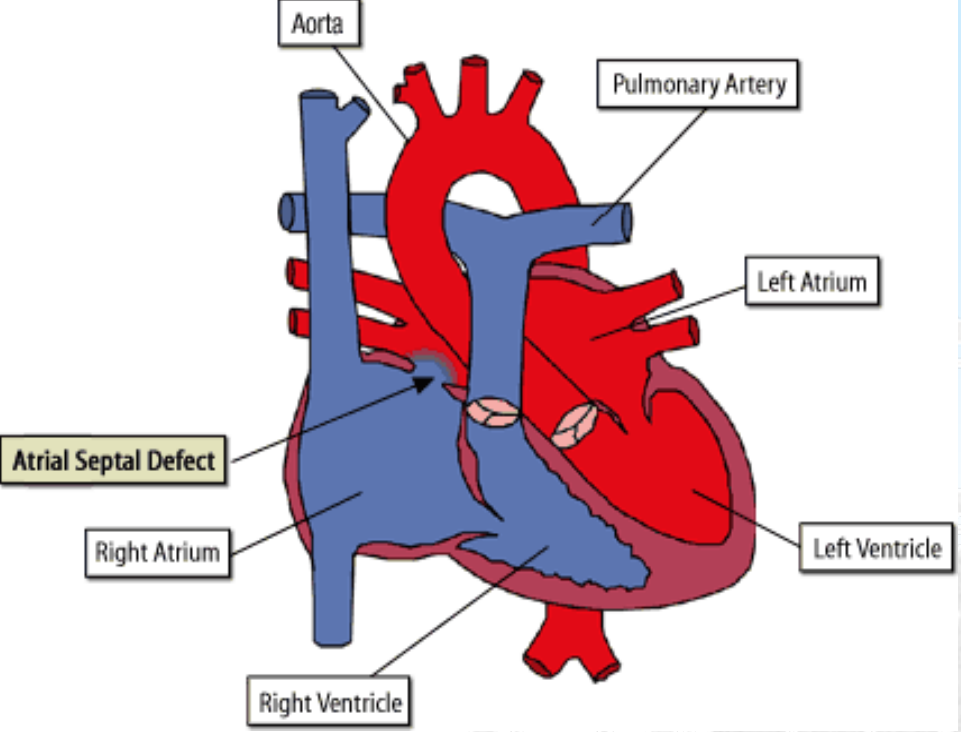
房间隔缺损

Atrial septal defect (ASD)

- The second common CHD
- 10-15% of CHD
- Female:male=2:1
- Single abnormality

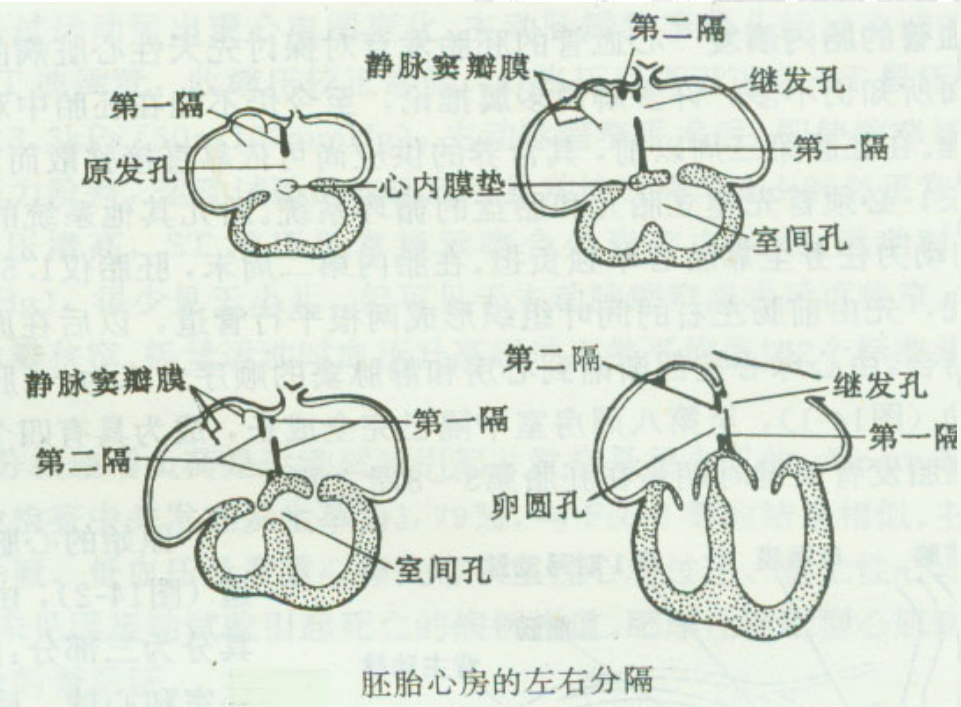
Associated lesions

associated with PAPVC and with abnormality of the atrioventicular valves(AVSD)

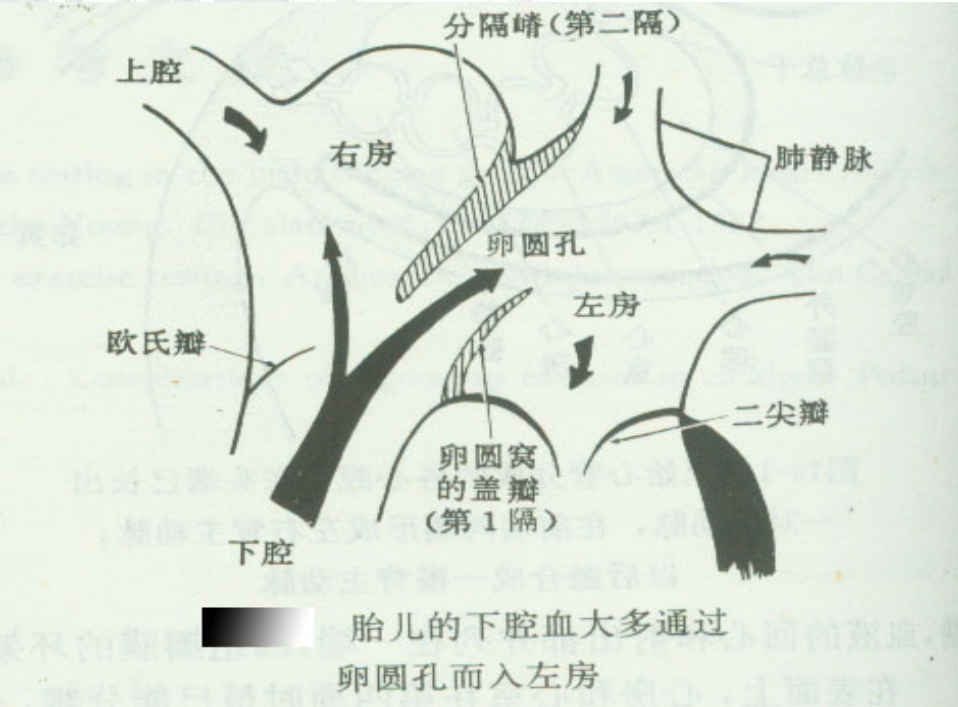


Anatomy of ASD

- **The ostium secundum type**
  (继发孔型房间隔缺损)
- **The ostium primum type**
  （原发孔型房间隔缺损）
- **Persistent foramen ovale type**
  （卵园孔型房间隔缺损）



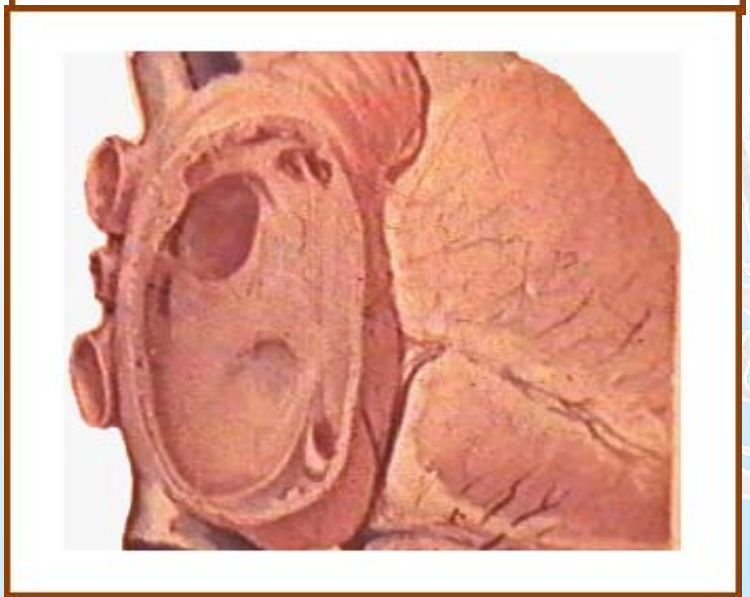
胚胎心房的左右分隔



胎儿的下腔血大多通过卵圆孔而入左房

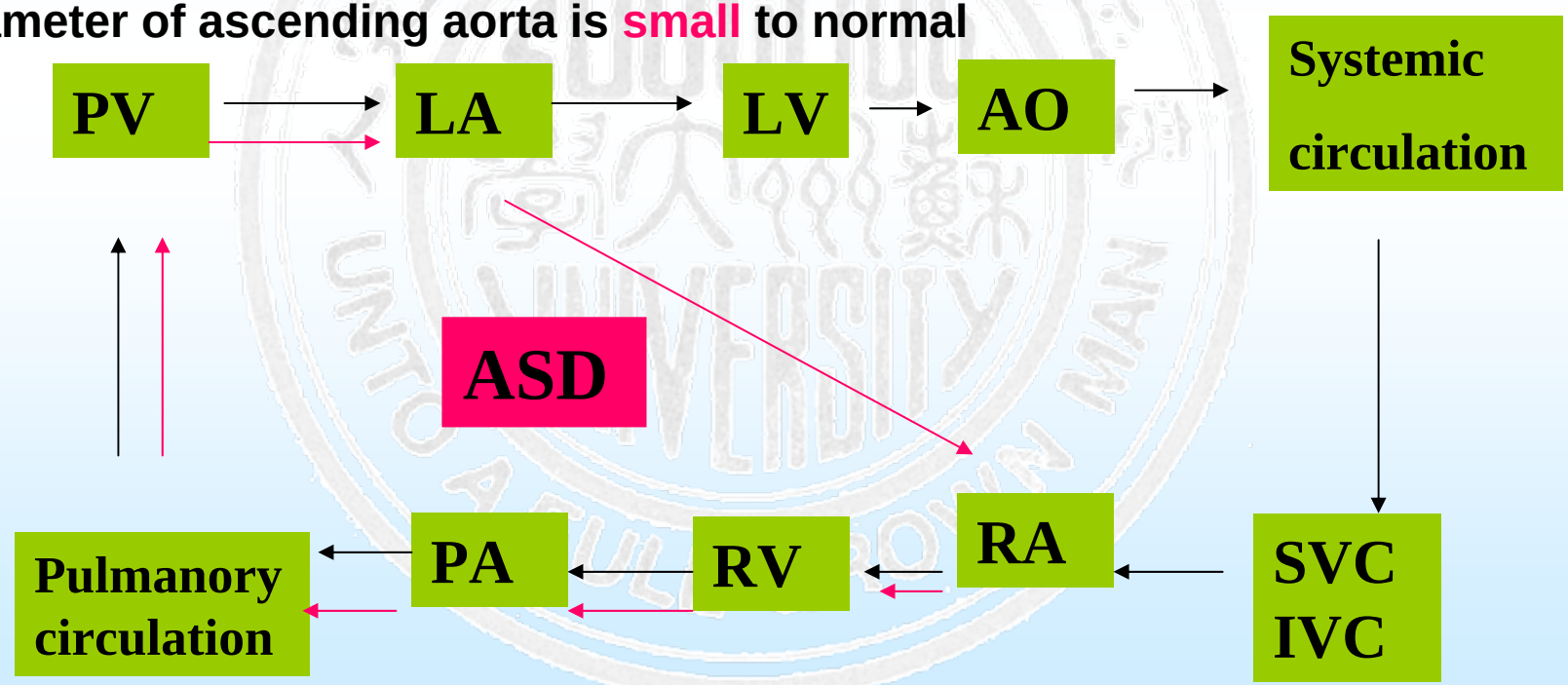
Types of ostium secundum ASD

继发孔房间隔缺损的解剖类型



Hemodynamics changes
血流动力学改变

- The blood in lung field **increased,** Blood in systemic circulation **decreased**
- **Pulmonary hypertension(PH) ,reversible -------- irreversible Eisenmenger syndrome**
- **Cardiac enlargement (RV,RA)**
- **Diameter of ascending aorta is small to normal**



Symptoms

(depending on the shunt of ASD)

small shunt ASD

- **No cardiovascular sign (most common)**

Moderate---large shunt ASD

- **Recurrent respiratory infections**
- **Difficulty with feeding, Poor weight gain**
- **Dyspnea**
- **Right sided heart failure**

Signs

- Pink (Acyanotic) ----cyanotic
- Parasternal (right ventricular) lift
- P_2 widely split and usually fixed (because of relatively pulmonary stenosis)

  P_2 accentuated (Result from pulmonary hypertension)
- ejection systolic murmur best heard third left intercostal space
- Mid-diastolic murmur at the lower left sternal border (if the shunt is significant in size)
- Arrythmias (in adulthood)
- Congestive heart failure (in adulthood)

Ⅱ－Ⅲ／Ⅵ级收缩期喷射性杂音

P2亢进伴固定分裂，喀喇音

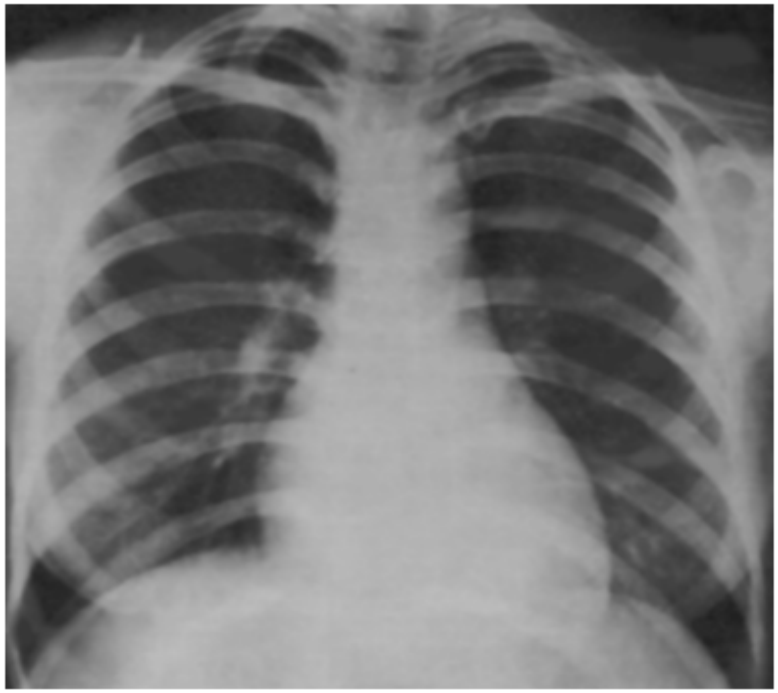
This murmur is caused by excessive flow across the pulmonary valve-- relatively pulmonary stenosis

Investigation and diagnosis

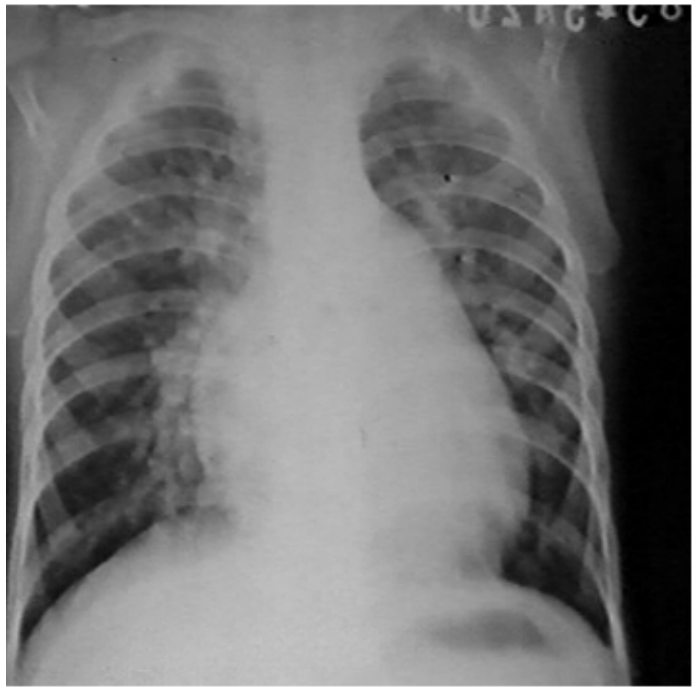
- Chest X-ray (CXR)
- Electrocardiography(ECG)
- Echocardiography(2DE)
- Cardiac catheterization and angiocardiography

Chest X-ray

- The pulmonary vascular markings increased
- The main pulmonary artery segment dilated
- Cardiac enlargement (RV,RA)
- Diameter of ascending aorta is small to normal

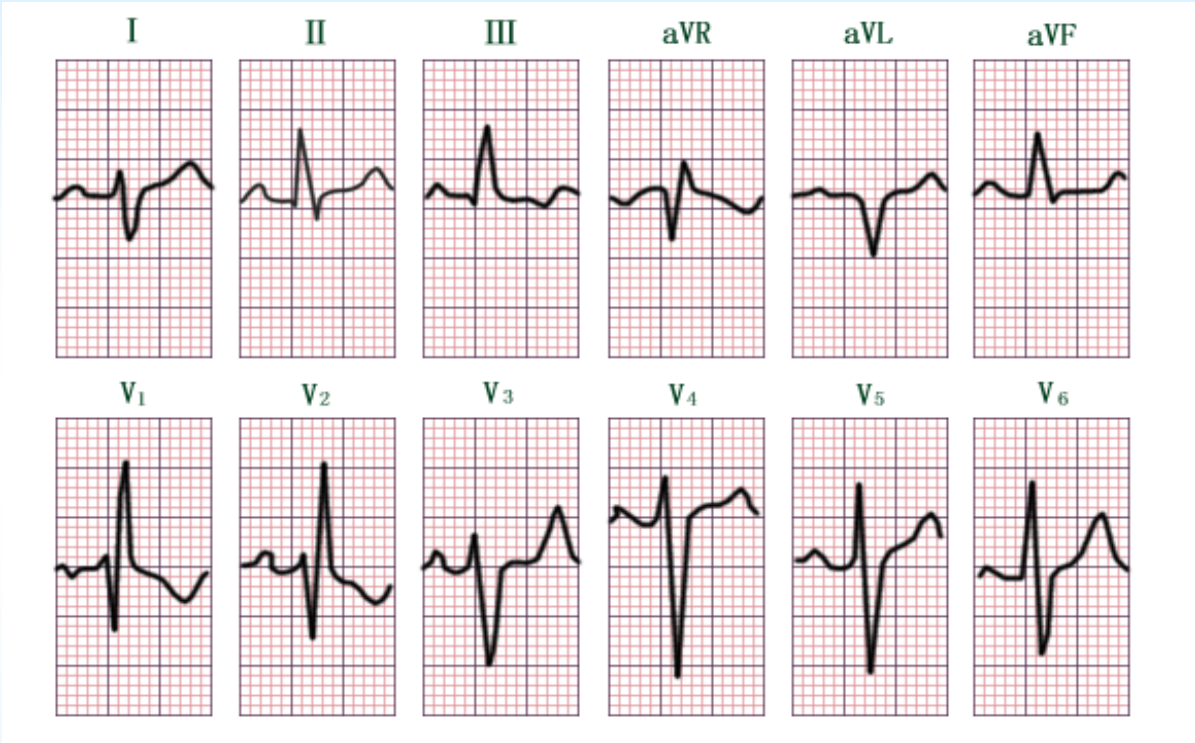


Normal



ASD

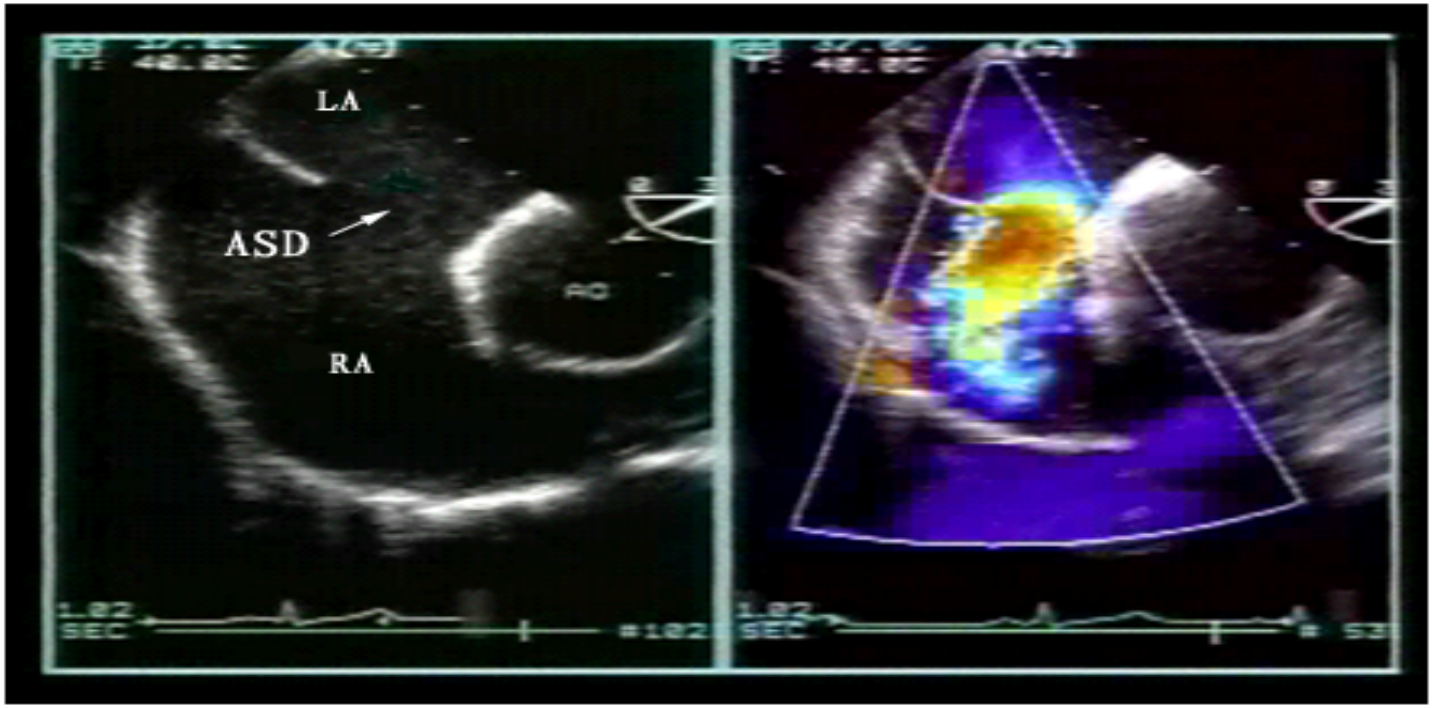
ECG of ASD

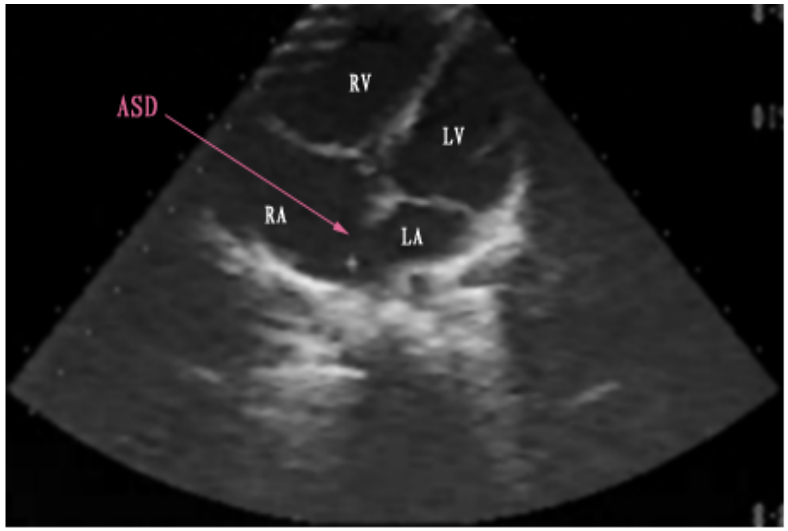


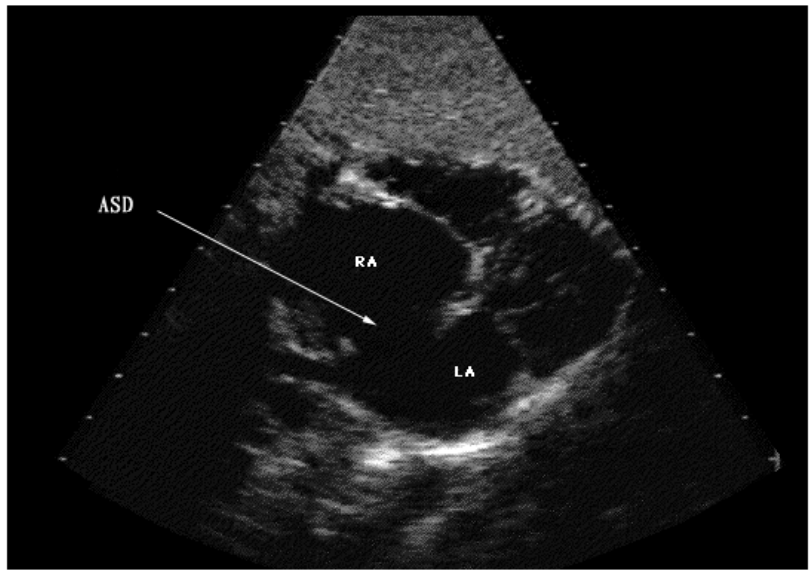
- Normal or right axis deviation (ostium secundum ASD)
- rsR1 in lead V1 (right bundle branch block)
- Right ventricular hypertrophy(sometimes)
- Left axis deviation (ostium primum ASD)

Echocardiography (two-dismensional Echo 2DE)

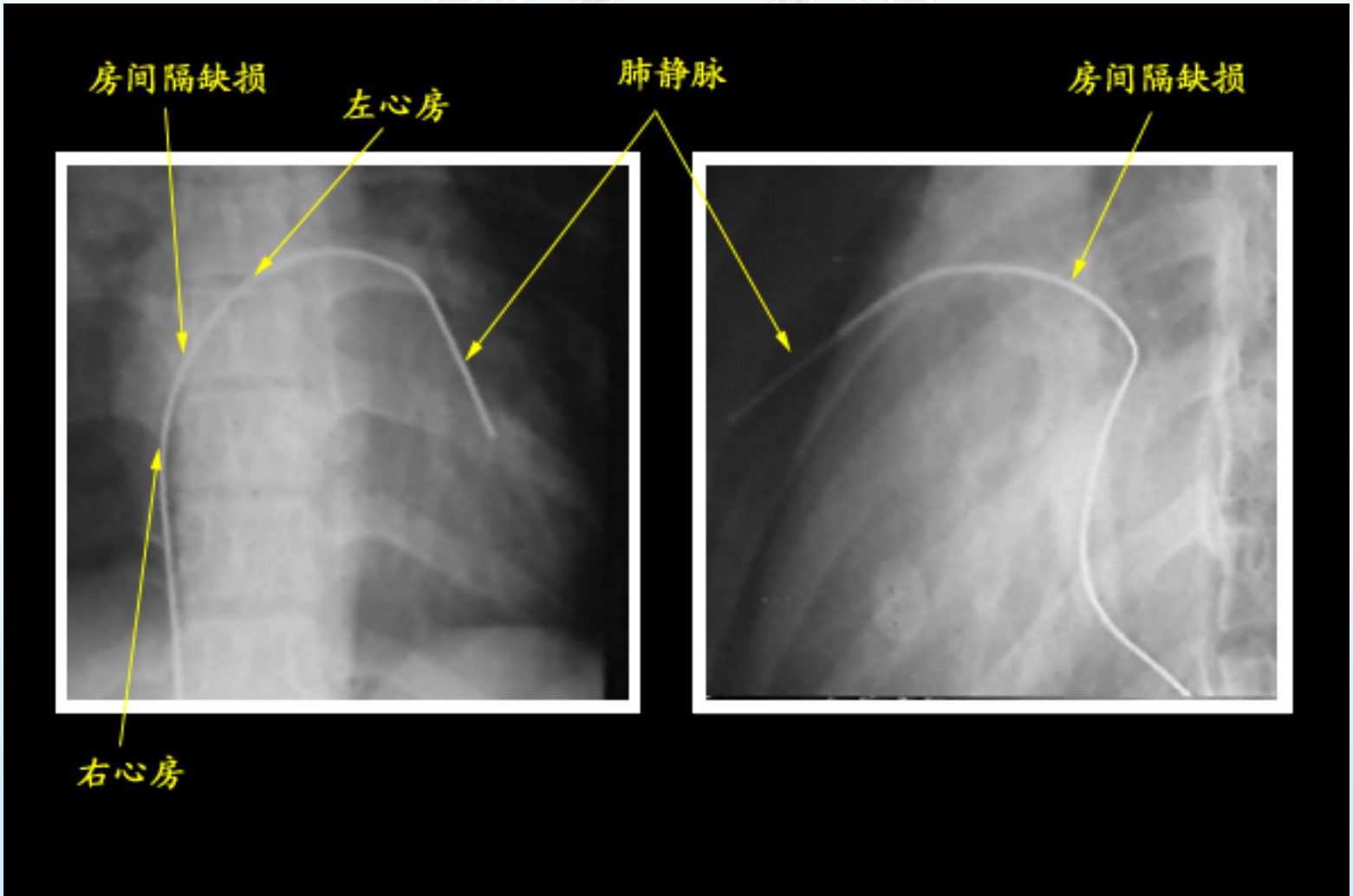
- The anatomic location
  (the number,size and location of ASD)
- Color flow doppler
  (the direction of the shunt)
- Estimate the pressure of pulmonary artery or pulmonary hypertension


LA
ASD
RA


ASD
RV
LV
RA
LA


ASD
RA
LA

Cardiac catheterization and angiocardiography



Course and prognosis

- Spontaneous closure (simple ASD)

(40%) age:< 1 y

- Adults with corrected defect

have a normal quality of life

Management

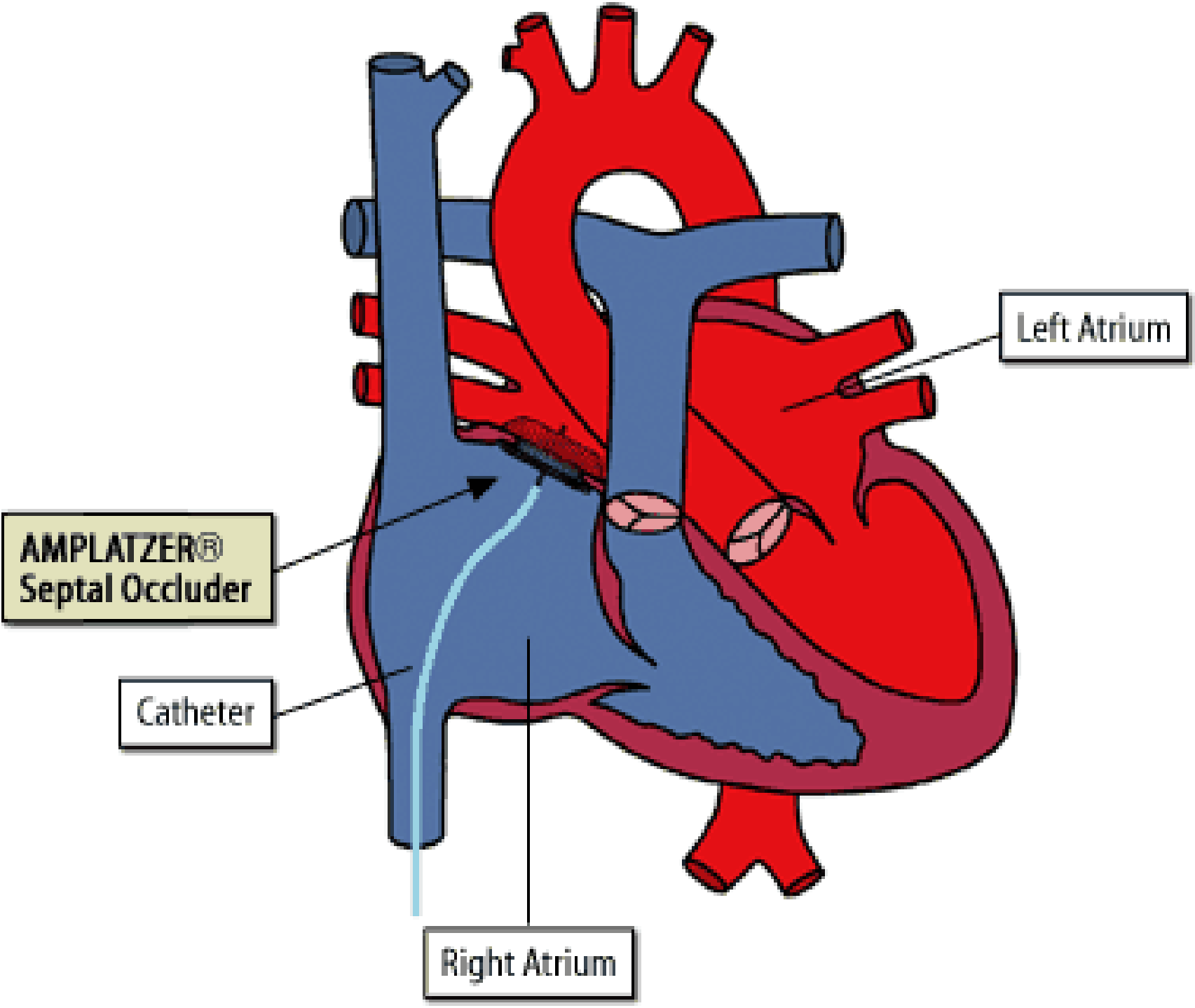
- **Management**

  Closure is recommended before school entry in all patients

- **Interventional therapy**

  Utilising transdefect clamping devices (e.g.Amplatzer occluder) have now become the first option for up to 80% of ASDs have a sufficient rim of tissue surrounding the defect

- **Surgical repair is necessary in 20%, involves direct suturing of small defects and the incorporadtion of an artificial patch (e.g.Dacron) in large defect**


Left Atrium
AMPLATZER®
Septal Occluder
Catheter
Right Atrium

Summary for ASD

- ASD, L—R shunt CHD,The symptom of ASD depends on the shunt, position and number of the defect.
- Respiratory infection is frequent with ASD patients
- The characteristic heart murmur and P2

1. I--III /Ⅵ ejection systolic murmur at the pulmonary area (L2--3)
2. Diastolic flow murmur at the left sternal border (if the shunt is significant in size)
3. P_2 widely split and usually fixed

Summary for ASD

- Enlarged chambers (RV ,or RA) can be detected by CXR ,ECG,and 2DE
- ECG with rsR^1 in lead V_1
- ASD can close spontaneously, especially in small size,under 1 y.
- Detecting PH is the key point in management of ASD patients

Ventricular Septal Defect (VSD)

室间隔缺损

Ventricular Septal Defect

- An abnormal interventricular connection .
- The most common CHD （occupy 30-50% of CHD）
- The defect may result from an incomplete fusion of the upper, membranous part of the septum (membranous VSD,75%)

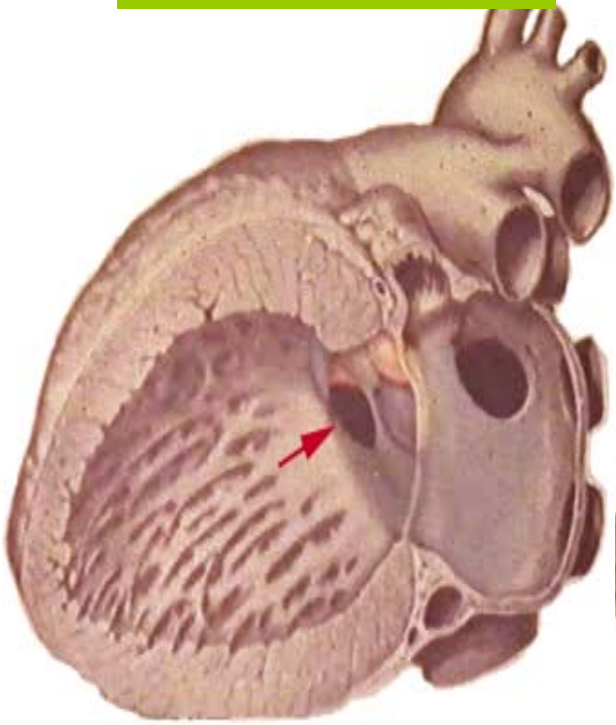
  Or the lower muscular septum(muscular VSD,20%)
- L—R shunt CHD （blood flows under pressure from the left to the right ventricle across the VSD）
- Simple abnormality/ associated lesion

Ventricular Septal Defect(VSD)

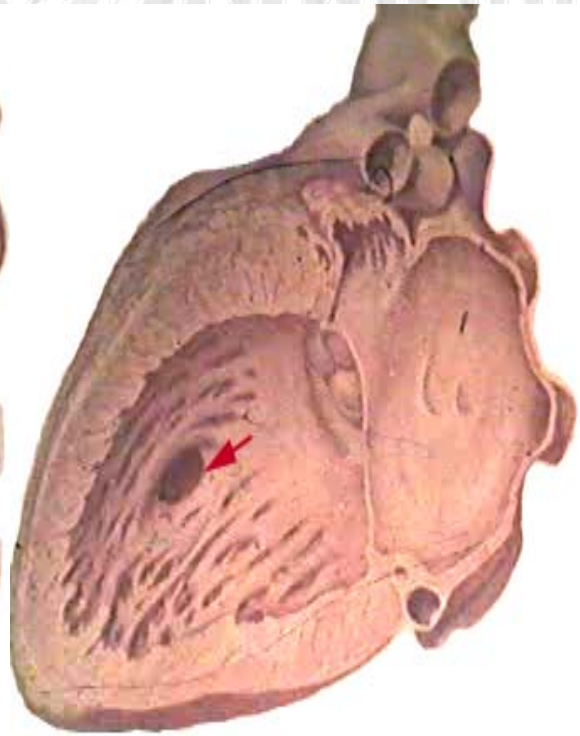
- Anatomy and Hemodynamics changes
- Clinical Finding (Symptoms and Signs)
- Investigation and Diagnosis
- Course and Prognosis
- Management and Treatment
- Summary and Questions

Anatomy of VSD

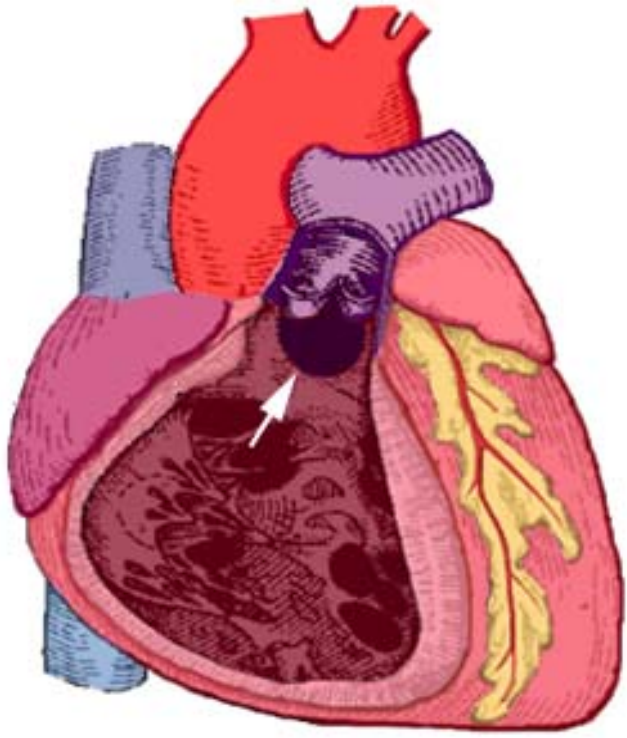
Membranous VSD 75%



muscular VSD (often multiple) 20%

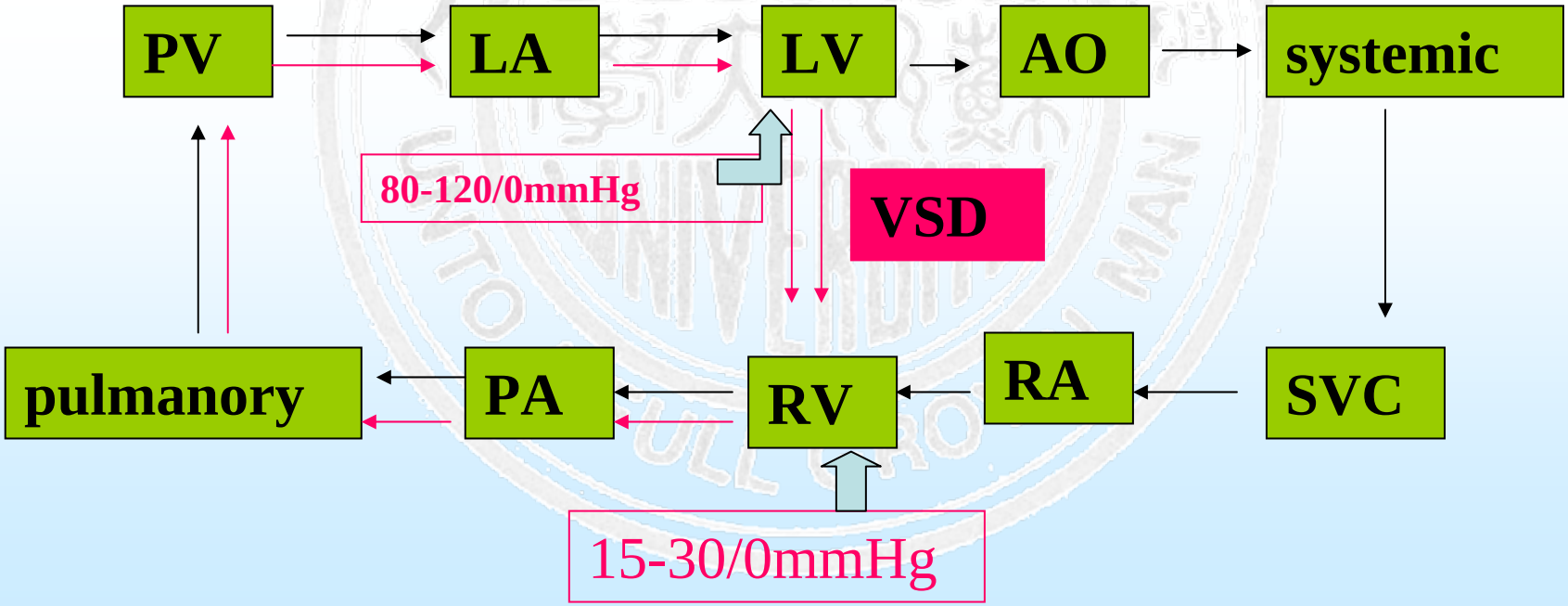


VSD below the pulmonary valve



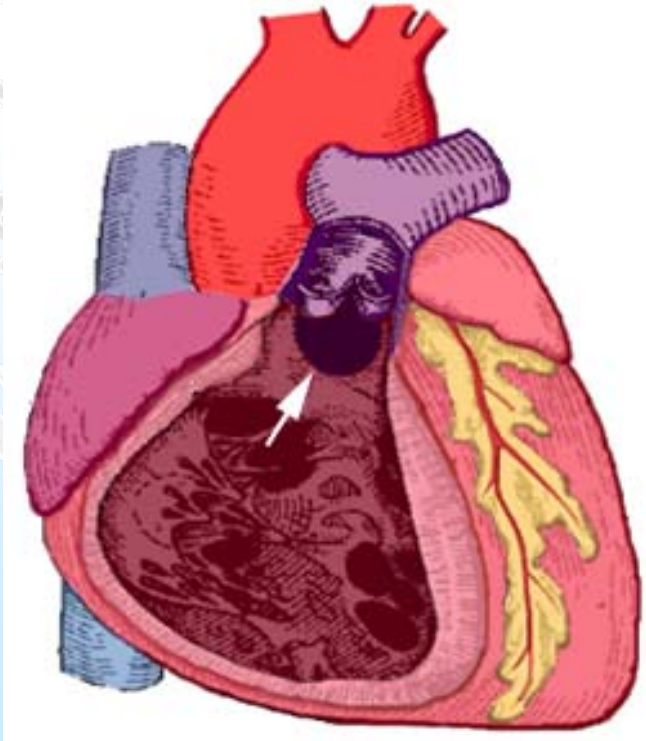
Hemodynamics changes

- The blood in lung field -----increased,
  Blood in systemic circulation ----decreased
- Pulmonary hypertension(PH) ,
  reversible -------- irreversible (Eisenmenger syndrome)
- Cardiac enlargement (LV,RV,LA)
- Diameter of ascending aorta is small to normal



Symptoms and signs of VSD

- depending on the shunt , size , number and the position of defects



Symptoms and signs of small VSD (<0.5cm^2)

- **Symptoms**

  General asymptomatic

- **Symptoms**

  Pink,normal pulse and BP

  A thrill may be present (sometimes)

  Normal heart sounds

  ESM (or PSM) at lower LSE

- **Investigations**

  ECG :normal

  CXR: normal

Symptoms and signs of moderate VSD (0.5—1.0cm^2)

- **Symptoms**

  Asymptom or recurrent chest infections

- **Symptoms**

  Pink, normal pulse and BP

  Thrill at LSE and LV impulse possible

  loud P2 best in the pulmonary area

  2-6/6 PSM all over precordium

  MDM

  pulmonary crepitation

  hepatomegly

- **Investigations**

  ECG :LV hypertrophy, slight / moderate cardiomegaly

  CXR: slight/moderate cardiomegaly ,pulmonary plethora

Symptoms and signs of large VSD (>1.0cm^2)

- **Symptoms**

  CCF , recurrent chest infections and failure to thrive

- **Symptoms**

  Pink, normal pulse and BP

  Apical beat forceful and displaced parasternal heave

  Thrill at LSE and LV impulse possible ,third heart sound

  loud P2 best in the pulmonary area

  2-6/6 PSM all over precordium

  MDM

  pulmonary crepitation / hepatomegly

- **Investigations**

  ECG : LV hypertrophy, RV hypertrophy

  CXR: Pulmonary plethora,cardiomegaly

Investigation and Diagnosis

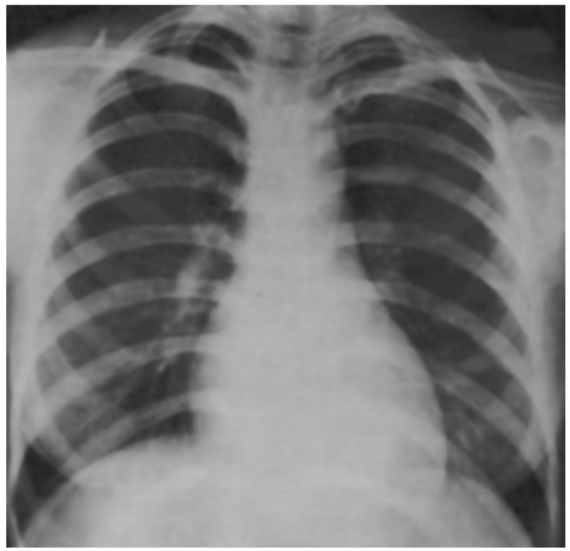
- Chest X-ray (CXR)
- Electrocardiography(ECG)
- Echocardiography(2DE)
- Cardiac catheterization and angiocardiography

Chest X-ray (CXR)

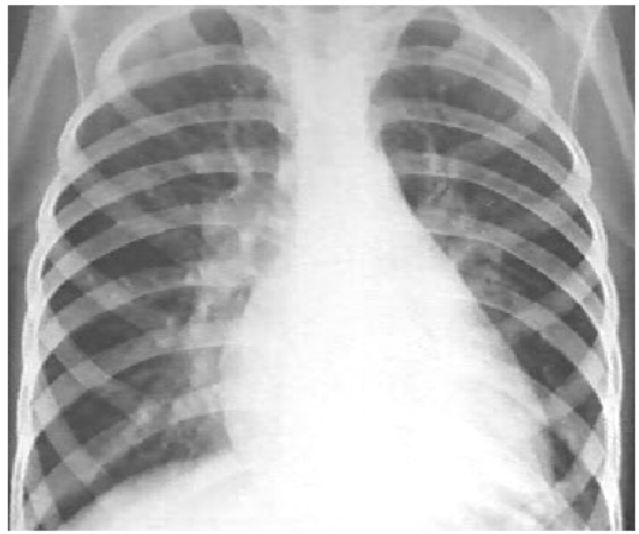
- The pulmonary vascular markings **increased**
- The main pulmonary artery segment **dilated**
- Cardiac enlargement **(LV,RV,LA)**
- Diameter of ascending aorta is **small** to normal

Chest X-ray

1. The pulmonary vascular markings increased
2. The main pulmonary artery segment dilated
3. Cardiac enlargement (LV,RV,LA)
4. Diameter of ascending aorta is small to normal



normal



abnormal

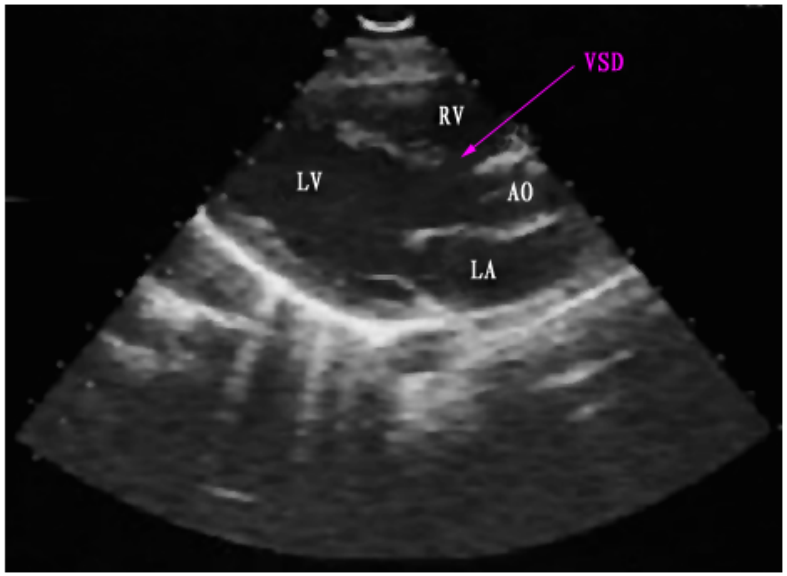
Electrocardiography (ECG)

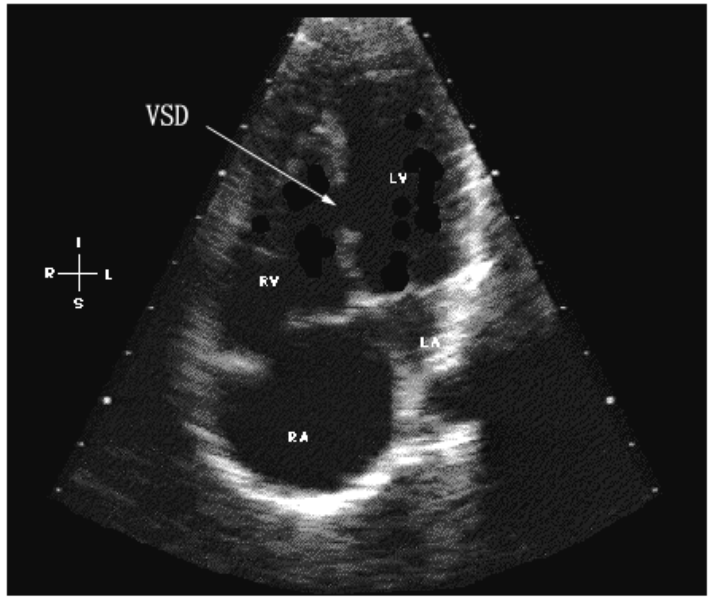
- Normal or left axis deviation
- Left ventricular hypertrophy
- LV and RV hypertrophy
- Pure RV hypertrophy

Echocardiography

1. The anatomic location (the number,size and location of VSD)
2. Color flow doppler (the direction of the shunt)
3. Estimate pressure

- The pressure of pulmonary artery (PH)
- Gradient between LV and RV


VSD
RV
LV
AO
LA


VSD
LV
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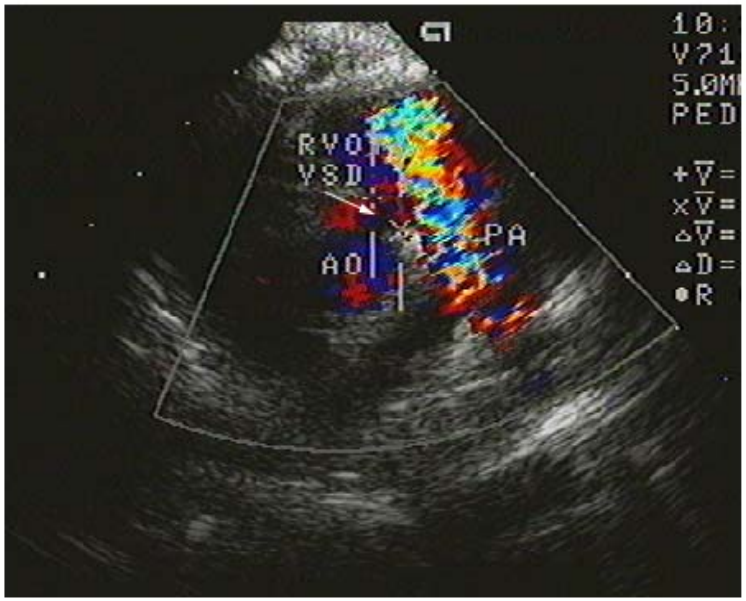
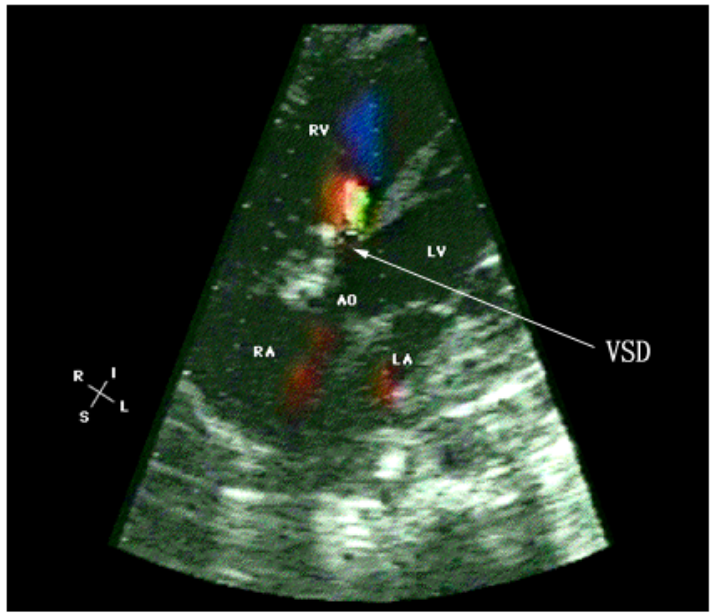

RVOT
VSD
AO
PA
+V=
xV=
△V=
△D=
●R


RV
LV
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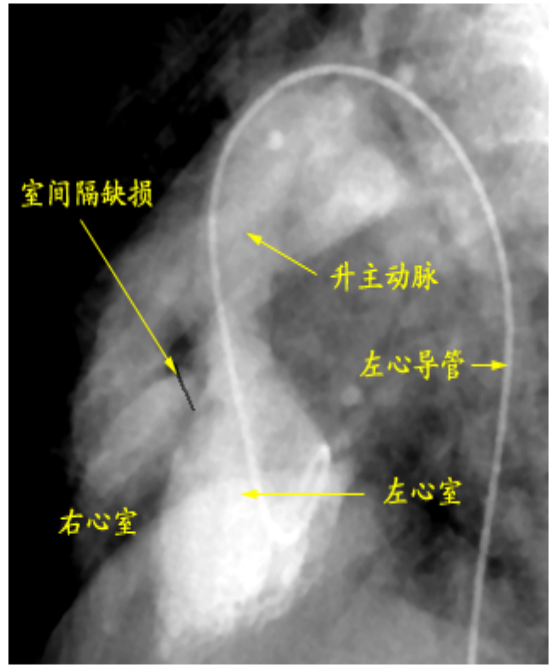
Cardiac catheterization and angiocardiography

- Diagnosis
- Estimate PH characteristic
- Intenventional therapy



Course and prognosis

- **Spontaneous closure (simple VSD)**

(25-40%) age:less than 1-5y

except defect just below the pulmonary valve.

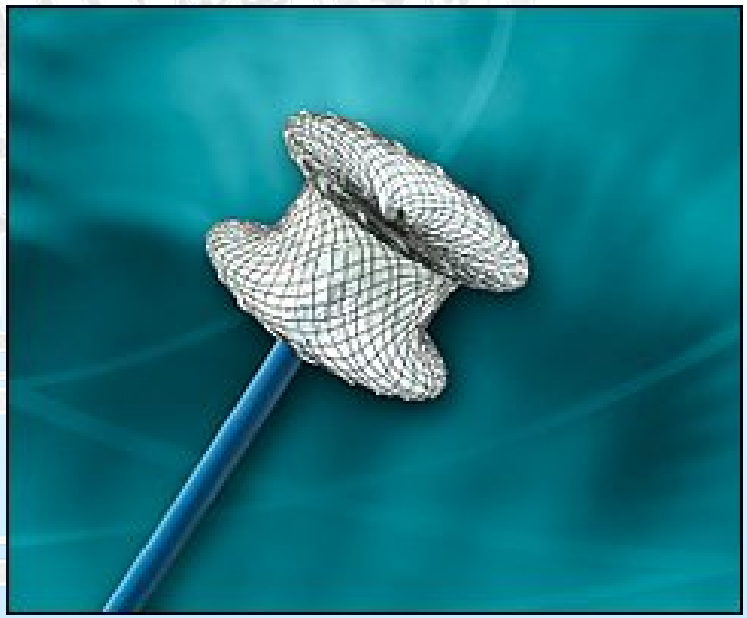
- **Adults with corrected defect**

have a normal quality of life

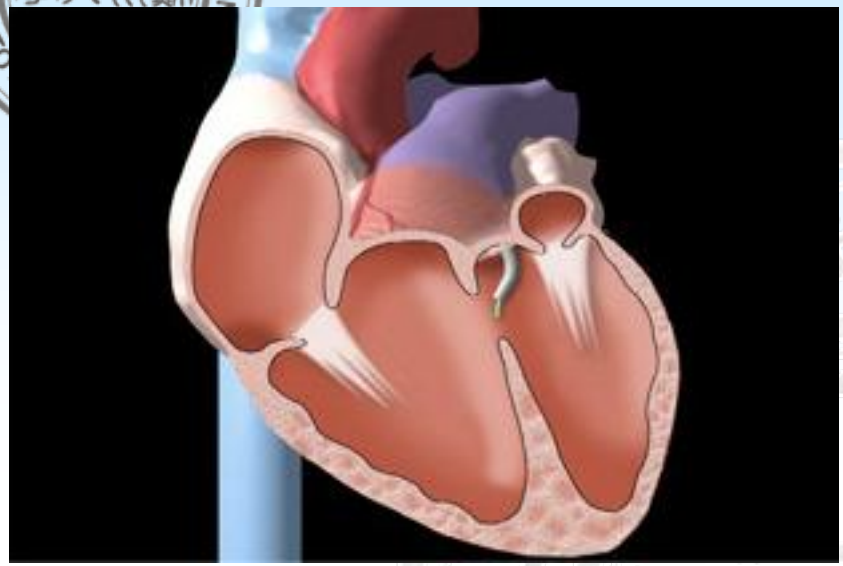
Management and Treatment

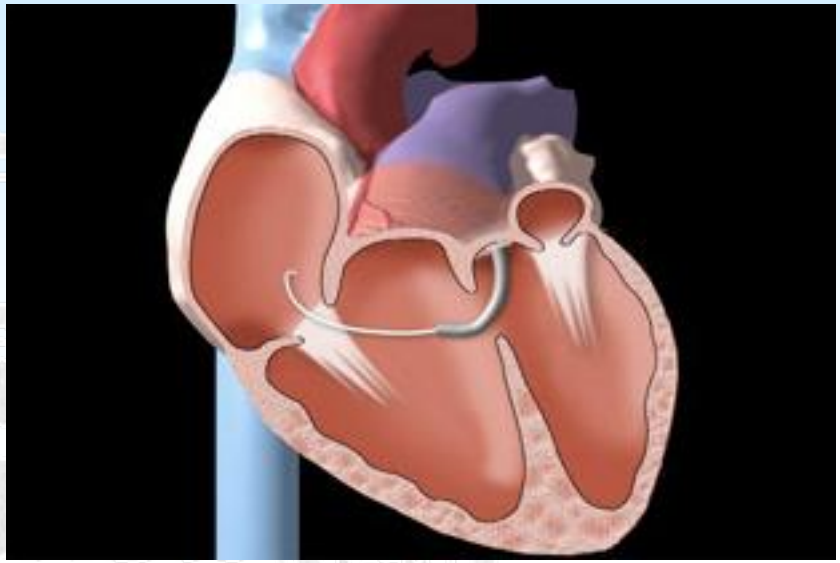
1. Medical management
2. Surgery
3. Interventional therapy

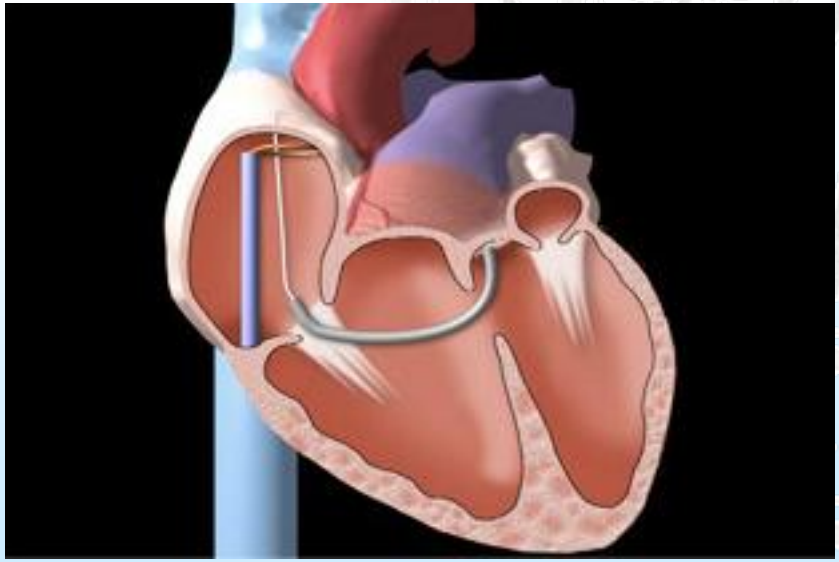
closure devices

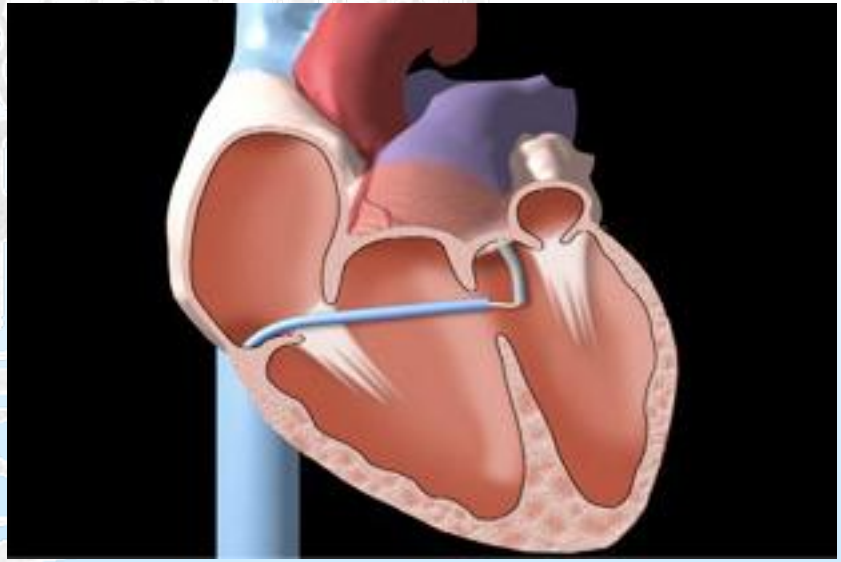


Steps for Interventional therapy









Summary

- VSD is the most common CHD, L—R shunt
- The symptom of VSD depends on the shunt, position and number of the defect.
- The characteristic heart murmur and P2
- Respiratory infection(pneumonia) and congestive heart failure are frequent with VSD patients

Summary

- VSD can close spontaneously, especially in small size,except defect below the pulmonary valve.
- Enlarged chambers (LV,RV ,or RA) can be detected by CXR ,ECG,and 2DE
- Detecting PH is the key point in management of VSD patients

Questions

1. How to estimate the PH in VSD patient in clinical experience? Why?
2. Important Concept:

   Pulmonary hypertension

   Roger's disease

   Eisenmeger syndrome

Thanks for your attention