ATRIAL SEPTAL DEFECT

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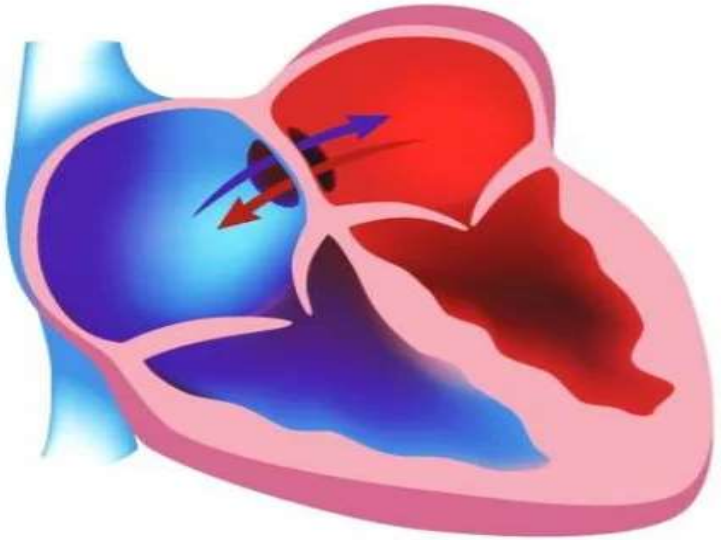
Introduction

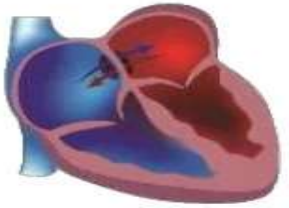
Atrial septal defect (ASD) is an abnormal communication between the atria due to a defect in the interatrial septum.



Incidence

- ASDs constitute 8% to 10% of congenital heart defects in children.
- The recent incidence of ASDs has been estimated to be 100 per 100,000 live births.
- The female:male ratio for secundum ASDs is 2:1, but for the sinus venosus ASDs it is 1:1 .

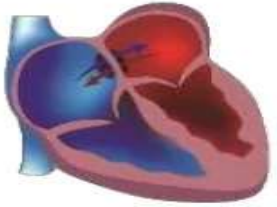
Moss and Adams' Heart disease in infants, children and adolescents, 9th edition



Incidence

About 30% to 50% of children with congenital heart defects have an ASD as part of the cardiac defect.

Park's Pediatric Cardiology for Practitioners, 6th edition



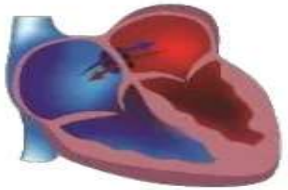
Epidemiology

The majority of cases of ASD are sporadic.

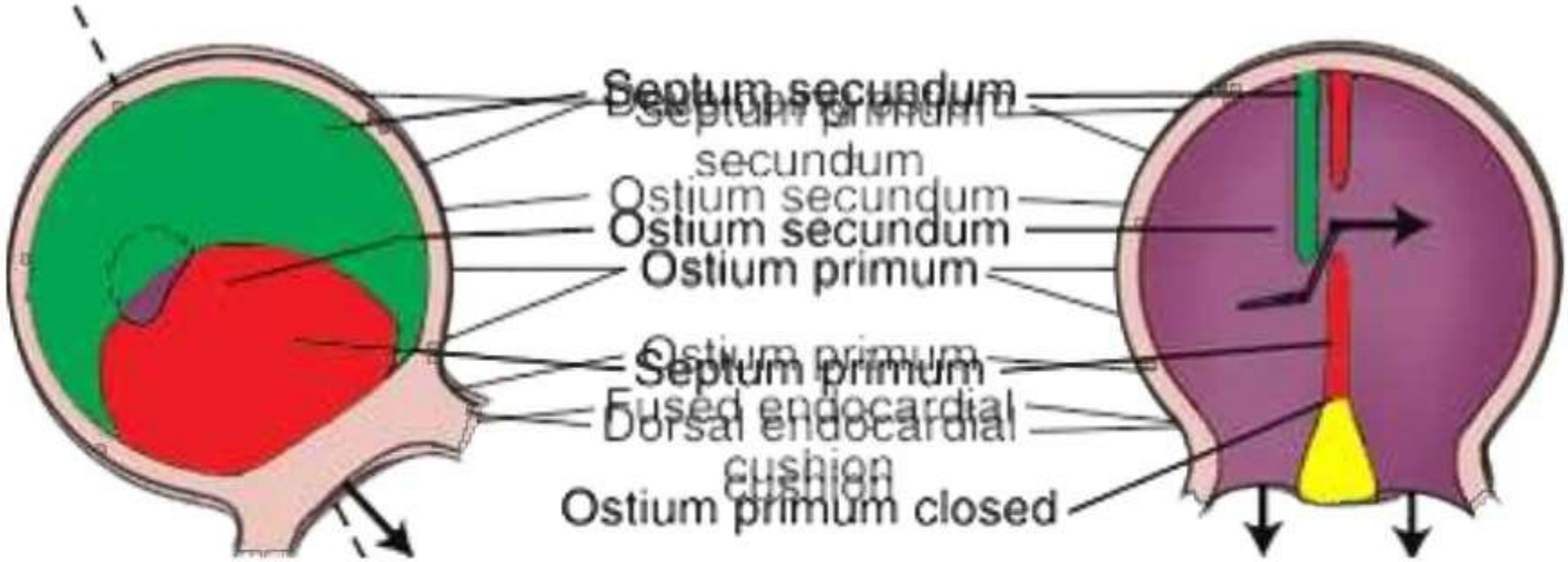
Autosomal dominant inheritance does occur as part of the

- Holt-Oram syndrome (hypoplastic or absent thumbs, radii, triphalangism, phocomelia, 1st-degree heart block, ASD) or in
- families with secundum ASD and heart block.

Nelson Textbook of Pediatrics, 20th edition



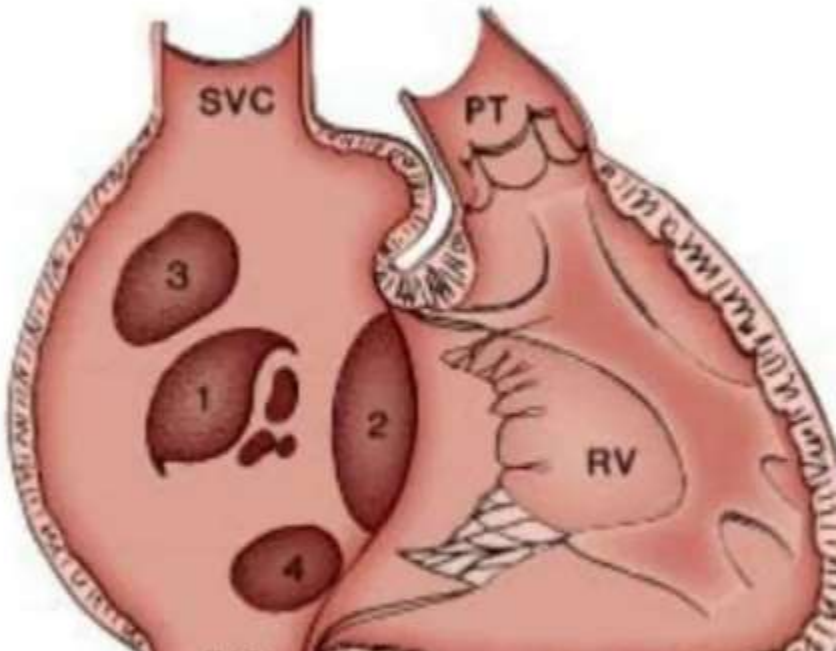
Formation of the Interatrial septum


Septum secundum
Septum primum
secundum
Ostium secundum
Ostium secundum
Ostium primum
Ostium primum
Septum primum
Fused endocardial
Dorsal endocardial
cushion
cushion
Ostium primum closed

Types of ASD

1. Secundum ASD (75%)
2. Ostium primum ASD (20%)
3. Sinus venosus ASD (5%)
4. Coronary sinus ASD (<1%)

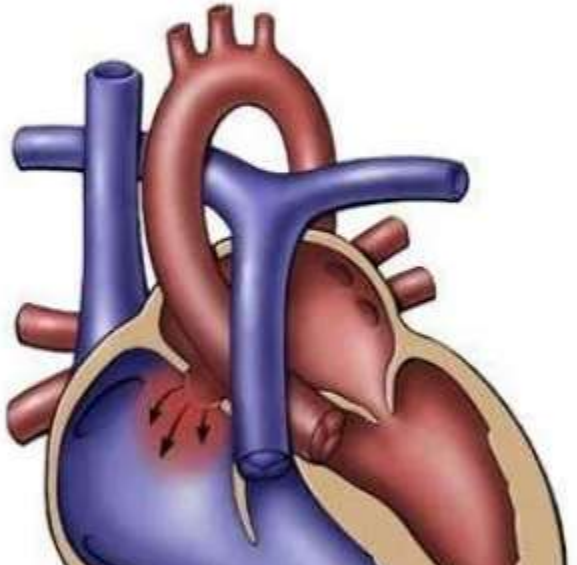
Moss and Adams' Heart disease in infants, children and adolescents, 9th edition



Secundum ASDs

Location:

- This defect is present at the site of fossa ovalis.
- Secundum ASDs may be single or multiple (fenestrated atrial septum).



Secundum ASDs (cont)

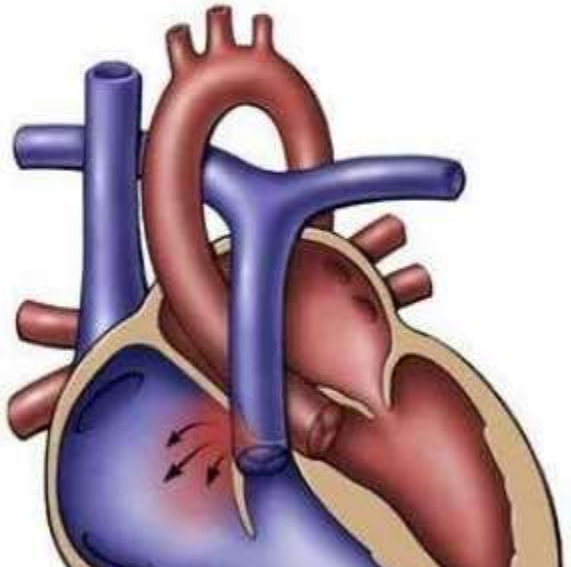
Associations:

- Partial anomalous pulmonary venous return
- Pulmonary valvular stenosis
- VSD
- Pulmonary artery branch stenosis
- Persistent left superior vena cava
- Mitral valve prolapse and insufficiency

Ostium primum ASD

Location:

- In the lower portion of the atrial septum and overlies the mitral and tricuspid valves.



Ostium primum ASD (cont)

Associations:

Isolated ostium primum ASD occurs in about 15% of all ASDs.

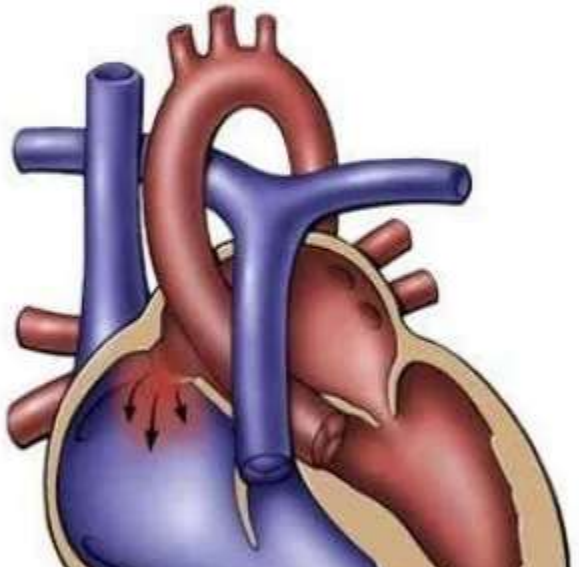
Other defects include-

- Ostium primum defects with clefts in the anterior mitral and septal tricuspid valve leaflets and
- Small ventricular septal defects.

Sinus venosus ASD

Location:

- It is most commonly located at the entry of the SVC into the RA (superior vena caval type) and
- rarely at the entry of the IVC into the RA (inferior vena caval type).



Sinus venosus ASD (cont)

Associations:

- The superior vena caval type is very commonly associated with anomalous drainage of the right upper pulmonary vein (into the RA), and
- The inferior vena caval type is often associated with anomalous drainage of the right lung into the IVC ("scimitar syndrome")

Coronary sinus ASD

Location:

- There is a defect in the roof of the coronary sinus, resulting direct communication with the left atrium.

Types:

- Complete unroofing of the CS
- Partial unroofing of the CS

Associations:

Pathophysiology of ASD

The degree of left to right shunting is dependent on:

- the size of the defect
- the relative compliance of the right ventricle and left ventricle and
- the relative vascular resistance in the pulmonary and systemic circulations.

Pathophysiology (cont)

In moderate-to-large defects, a considerable shunt of oxygenated blood flows from the left to the right atrium

↓

Volume overload and dilation of the right atrium and ventricle

↓

The tricuspid and pulmonary annuli may dilate and become incompetent

↓

Pathophysiology (cont)

Pulmonary arteries, capillaries & the veins are dilated

↓

Flow-related pulmonary artery hypertension

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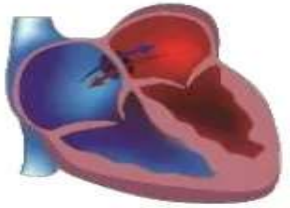
Medial hypertrophy of pulmonary arteries and muscularization of the arterioles resulting in pulmonary vascular obstructive disease

↓

Reversal of the shunt

↓

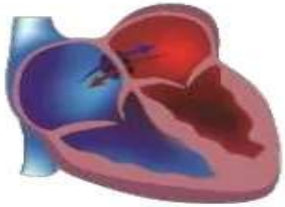
Eisenmenger syndrome



Clinical Presentation

History

- Asymptomatic
- Infants
  - features of pulmonary overcirculation
  - recurrent respiratory infections
  - failure to thrive
- Older children
  - mild fatigue
  - dyspnea that may worsen with age



Physical Examination

Examination of precordium

Inspection:

Left precordial bulge.

Palpation:

Prominent right ventricular impulse felt along the lower left sternal border and the subcostal area.

Physical Examination (cont)

Auscultation:

- Wide, fixed splitting of the second heart sound (S2).
- A systolic ejection murmur best heard at the left upper sternal border.
- A short, rumbling mid-diastolic murmur is often audible at the lower left sternal border.

Diagnostic Evaluation

Investigations

- Chest X-RAY
- ECG
- Echocardiography
- Cardiac catheterization (occasionally)

Chest X-Ray

A small shunt across the ASD

- normal-appearing CXR

Patients with significant shunts

- Cardiomegaly, due to right atrial and right ventricular enlargement, and
- Increased pulmonary vascular markings extending

Electrocardiogram

A small left-to-right shunt and no right atrial or ventricular dilation, the ECG is normal.

Secundum ASD

A significant left-to-right shunt

- Right axis deviation
- Right ventricular hypertrophy
- rsR’ pattern in V1

Electrocardiogram (cont)

Ostium Primum ASD

- Left axis deviation

Echocardiography

Two-dimensional echo study is diagnostic.

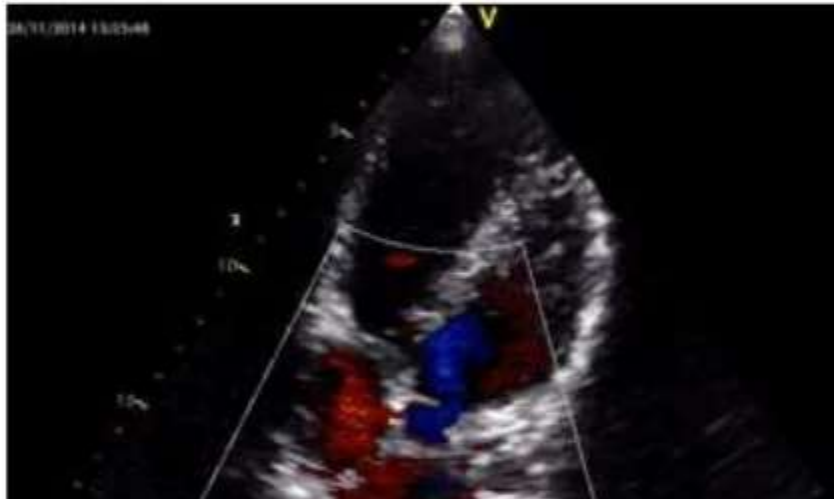
It shows

- the position as well as the size of the defect.
- the enlarged right atrium, right ventricle, and pulmonary arteries.
- associated anomalies such as

Echocardiography (cont)

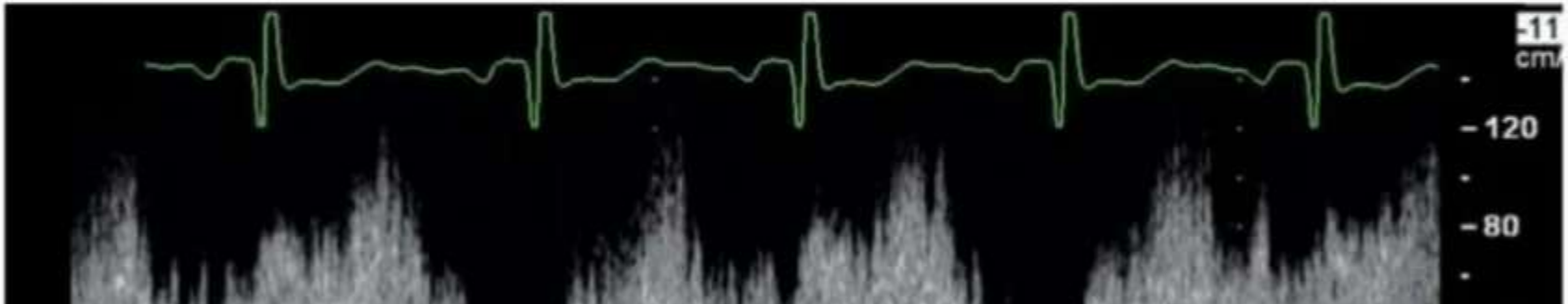
Color Doppler can visualize the shunt across the ASD.

- A qualitative assessment of the shunt across the ASD is done by direct visualization of the shunt and its effect on the right-sided cardiac chambers.



Echocardiography (cont)

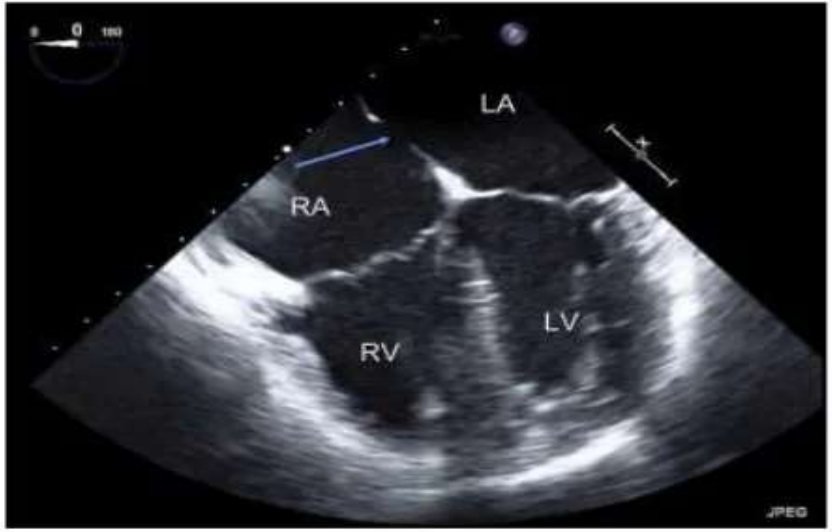
Pulsed Doppler examination reveals a characteristic flow pattern with the maximum left-to-right shunt occurring in diastole.



Echocardiography (cont)

Transesophageal echocardiography (TEE)

- may be used as an alternative for older children and adolescents, especially in those who are overweight.
- used to detect a sinus venosus ASD.
- is a monitoring adjunct for operative and per-cutaneous closure of ASD.



Echocardiography (cont)

Three-Dimensional Echocardiography

- allows better morphologic delineation of the ASD and its surrounding structures.
- used for guiding device closure during the procedure.

Cardiac Catheterization

Indicated

- If pulmonary vascular disease is suspected.
- Diagnosing associated lesions such as
  - ✓partial anomalous pulmonary venous return or
  - ✓mitral stenosis.
- Direct measurement of intracardiac and pulmonary artery pressure can be performed.

Management

Medical

1. Exercise restriction is unnecessary.
2. Asymptomatic infant and children are followed up at yearly interval until surgery is recommended prior to school entry.
3. Symptomatic, anti-congestive therapy with diuretics may be indicated until closure is accomplished.

Management (cont)

Closure of an ASD is indicated if there is a large shunt.

Indicators of a large shunt include

- Qp:Qs ≥ 1.5.
- A diastolic flow rumble in the tricuspid area,
- ECG evidence of right ventricular hypertrophy,
- Chest x-ray evidence of cardiomegaly or increased pulmonary vascular markings, or
- Echocardiographic evidence of right ventricular

Nonsurgical Closure

- Several closure devices that can be delivered through cardiac catheters have been shown to be safe and efficacious for secundum ASD closure.
- Currently there is no transcatheter devices designed for closer of sinus venosus, primum or coronary sinus ASD.

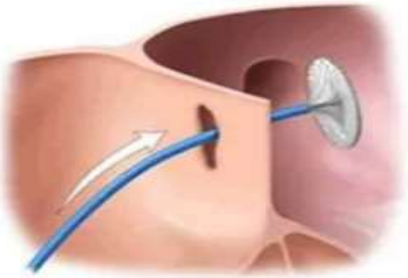
Nonsurgical Closure

Devices available for clinical use:

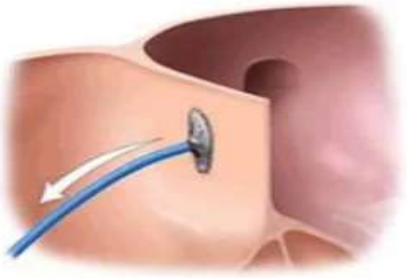
A. Amplatzer septal occlude
B. Gore Helix septal occlude
C. CardioSEAL device
D. BioSTAR device



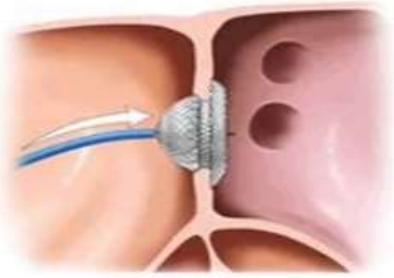

Nonsurgical Closure



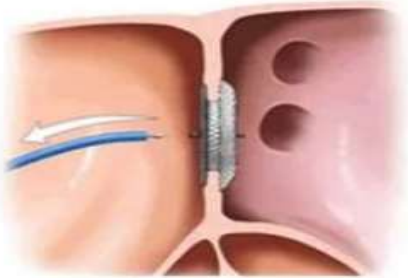
Step 1



Step 2



Step 3



Step 4

Nonsurgical Closure

Advantages

- Complete avoidance of cardiopulmonary bypass.
- Avoidance of pain and residual thoracotomy scars.
- A less than 24-hour hospital stay, and rapid recovery.

Nonsurgical Closure

Post-device closure follow-up:

The patients are administered aspirin 5 mg/kg/day for 6 months.

Post procedure echo studies check for

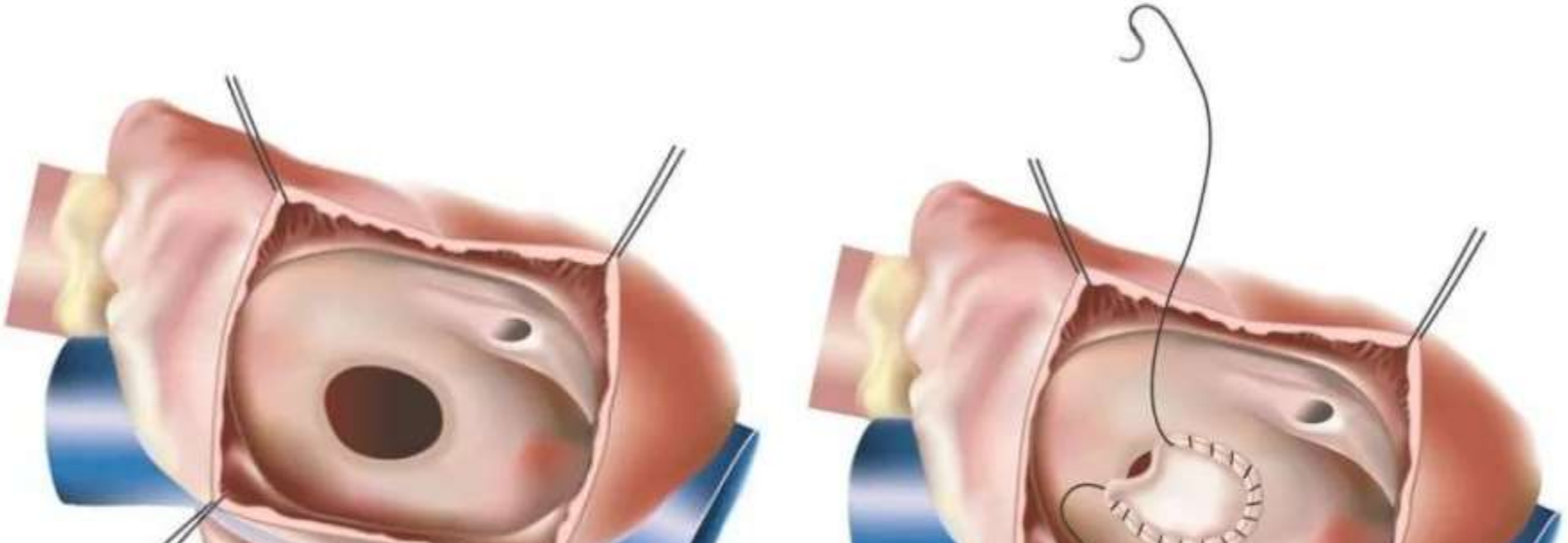
- a residual atrial shunt and
- unobstructed flow of pulmonary veins, coronary sinus, and venae cavae and
- proper function of the mitral and tricuspid valves

Surgical Closure

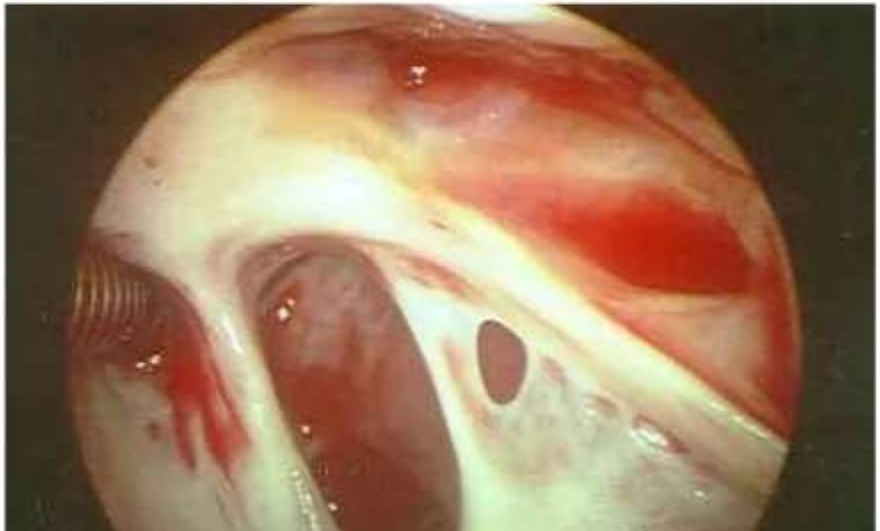
Indications and Timing

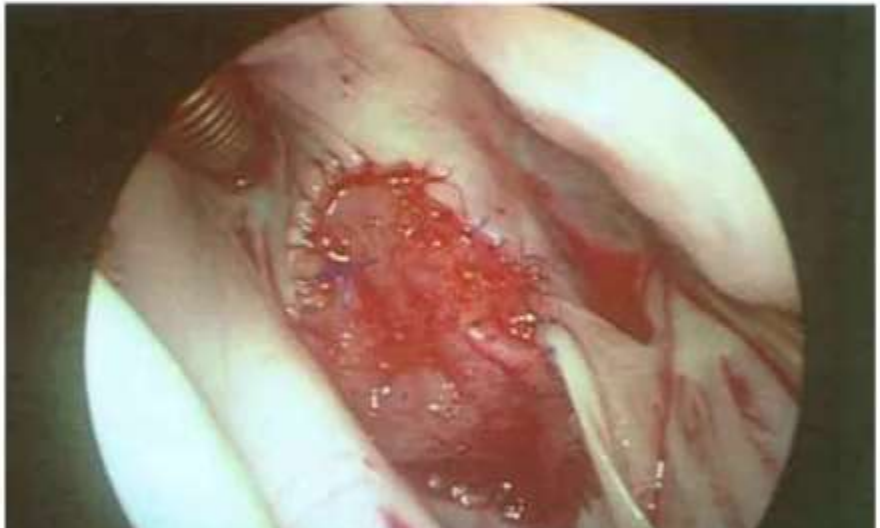
- A left-to-right shunt with a pulmonary-to-systemic blood flow ratio (Qp/ Qs) of ≥1.5:1 only if device closure is not considered appropriate.
- If CHF does not respond to medical management, surgery is performed during infancy, again if device closure is considered inappropriate.
- If oxygen and other medical therapy are needed for infants with associated bronchopulmonary dysplasia and the

Surgical Closure



Surgical Closure





Surgical Closure

Complications:

- Cerebrovascular accident and
- Postoperative arrhythmias

Surgical Closure

Postoperative Follow-up

- Cardiomegaly on x-ray film and enlarged RV dimension on echo as well as the wide splitting of the S2 may persist for 1 or 2 years postoperatively. The ECG typically demonstrates RBBB (or RV conduction disturbance).
- Atrial or nodal arrhythmias occur in 7% to 20% of postoperative patients. Occasionally, sick sinus syndrome, which occurs especially after the repair of a sinus venosus defect, may require antiarrhythmic drugs, pacemaker

Natural History of ASDs

- In patients with an ASD **less than 3 mm** in size diagnosed before 3 months of age, spontaneous closure occurs in 100% of patients at 1½ years of age.
- Spontaneous closure occurs more than 80% in patients with defects between **3 and 8 mm** before 1½ years of age.
- An ASD with a diameter **greater than 8 mm** rarely closes spontaneously.

Natural History of ASDs

- Most children with an ASD remain active and asymptomatic. Rarely, congestive heart failure (CHF) can develop in infancy.
- If a large defect is untreated, CHF 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.
- Infective endocarditis does not occur in patients with


Thank you