NOTES
ACYANOTIC DEFECTS

GENERALLY, WHAT ARE THEY?

PATHOLOGY & CAUSES

- Heart defects presenting without cyanosis (blue-tinged skin)
- Caused by fetal heart malformation, can lead to heart failure
- ASD, PDA, and VSD
  - All three cause left-to right shunt → oxygenated blood flows redundantly through pulmonary circulation → becomes Eisenmenger syndrome over time

SIGNS & SYMPTOMS

- Sometimes asymptomatic, but can lead to heart failure, Eisenmenger syndrome

Heart failure

- *Infants:* poor feeding/failure to thrive, fluid retention, pulmonary congestion, hepatomegaly, respiratory distress, elevated jugular venous pressure

Eisenmenger syndrome

- *At rest:* asymptomatic
- *With exertion:* cyanosis, palpitations dyspnea, chest pain, syncope

DIAGNOSIS

DIAGNOSTIC IMAGING

- Heart imaging to identify defect type

TREATMENT

SURGERY

- Rarely

MNEMONIC: P(C)AV

Acyanotic defects

Patent ductus arteriosus

(**C**oarctation of the aorta): no shunt

Atrial septal defect

Ventricular septal defect

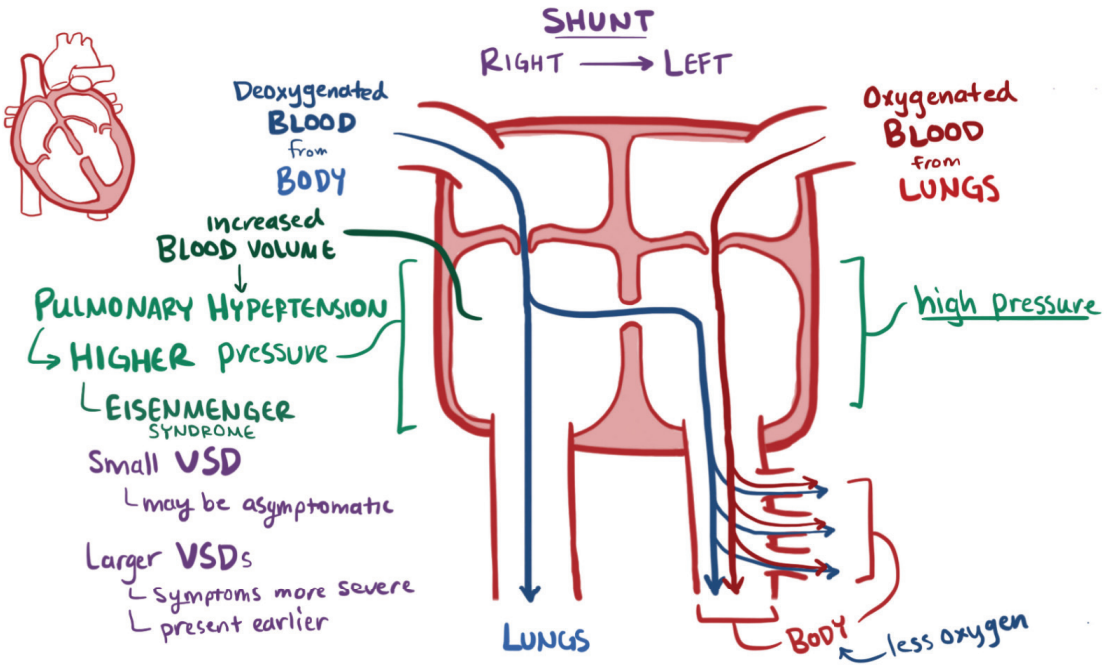

Figure 2.1 Illustration of blood flow through a ventricular septal defect.

ATRIAL SEPTAL DEFECT (ASD)

osms.it/atrial-septal-defect

PATHOLOGY & CAUSES

- A hole in the heart wall dividing left/right atria (left-to-right shunt)
- Blood passes through pulmonary circulation redundantly

SIGNS & SYMPTOMS

- Fixed, split S2 and pulmonic ejection murmur (louder with age)
- Infants and children
  - Respiratory infections
  - Failure to thrive
- Adults (before 40)
  - Palpitations, exercise intolerance, dyspnea, fatigue

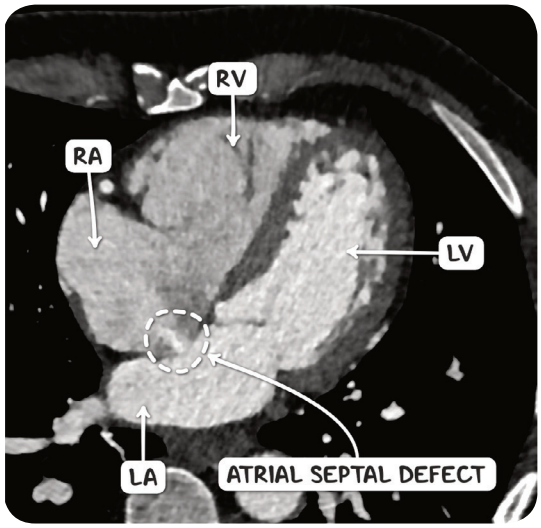

Figure 2.2 CT scan in the axial plane showing an atrial septal defect. Note the faint contrast plume as blood flows from the high pressure left system to the low pressure right system. RA; right atrium. LA; left atrium. RV; right ventricle. LV; left ventricle.

DIAGNOSIS

DIAGNOSTIC IMAGING

Chest X-ray

- Right heart dilation
- Prominent pulmonary vascularity

Transesophageal echocardiography

- Visualize size & location accurately

SURGERY

Right heart catheterization

- Increased oxygen saturation in:
  - Right atrium
  - Right ventricle
  - Pulmonary artery

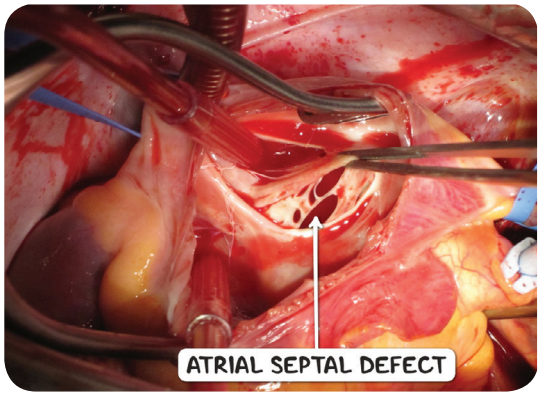

Figure 2.3 Intraoperative view of multiple, pinhole atrial septal defects.

TREATMENT

SURGERY

- Percutaneous surgical closure
- *Adults:* surgery in cases of
  - Right ventricular enlargement, paradoxical embolism, right-to-left shunt

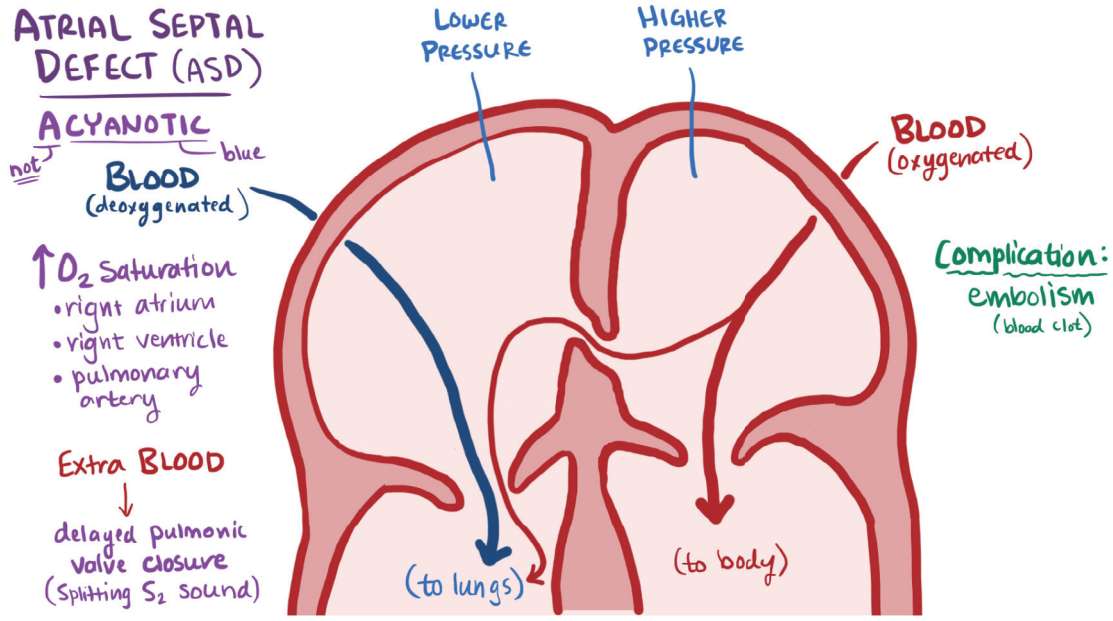

Figure 2.4 Illustration depecting blood shunting from left to right atrium in atrial septal defect.

COARCTATION OF THE AORTA (CoA)

osms.it/coarctation-of-the-aorta

PATHOLOGY & CAUSES

- Narrowed segment of aorta
- Upstream issues
  - Blood flow increases into aortic branches before coarctation → blood flow, pressure increases in upper extremities, head
- Downstream issues
  - Decreased blood flow, decreased pressure in lower extremities
  - Kidneys receive less blood → activate renin-angiotensin-aldosterone system (RAAS) → secondary hypertension
- Preductal coarctation
  - Associated with Turner syndrome, PDA
  - May go unnoticed unless severe. Presents as postductal coarctation
- Postductal coarctation
  - Distal to ligamentum arteriosum
  - Presents in adulthood
  - Blood pressure higher upstream, lower downstream
  - Autoregulatory vasoconstriction/ vasodilation preserves regional blood flow

SIGNS & SYMPTOMS

- Depends on presence/severity of PDA
- Systolic murmur
  - *Systole:* diamond-shaped murmur
  - *Diastole:* high-pitched decrescendo murmur

Infants

- Lower extremity cyanosis
- Absent or delayed femoral pulse
- Failure to thrive/poor feeding
- Blood pressure higher in upper extremities compared to lower extremities
- Secondary hypertension
- Severe heart failure, shock if/when PDA closes
- Other symptoms may more apparent with age
  - Chest pain, cold extremities, claudication on exertion
  - Left ventricular impulse palpable, sustained
  - Pulsations felt in intercostal spaces

Adults

- Hypertension (most common)
- Hypotension in lower extremities
- Bilateral lower extremity claudication
- Dyspnea on exertion
- Delayed/weak femoral pulses

DIAGNOSIS

DIAGNOSTIC IMAGING

Angiogram

- Visualize narrowing in aorta, anatomy & severity

Chest X-ray

- *Rib notching:* 3-sign (narrowed aorta resembles notch of number 3 due to prestenotic of aortic arch & postenotic of descending aorta dilatation)

Echocardiograph

- Visualize location, size, blood turbulance

OTHER DIAGNOSTICS

ECG

- Left ventricular hypertrophy, left atrial enlargement

TREATMENT

MEDICATIONS

Prostaglandin E

- Increases flow to lower extremities

SURGERY

- Resection with end-to-end anastomosis
  - If unfeasible, bypass graft across area of coarctation
- *Long-segment coarctation:* subclavian aortoplasty
- Prosthetic patch aortoplasty (rarely)
- Balloon angioplasty with possible stent

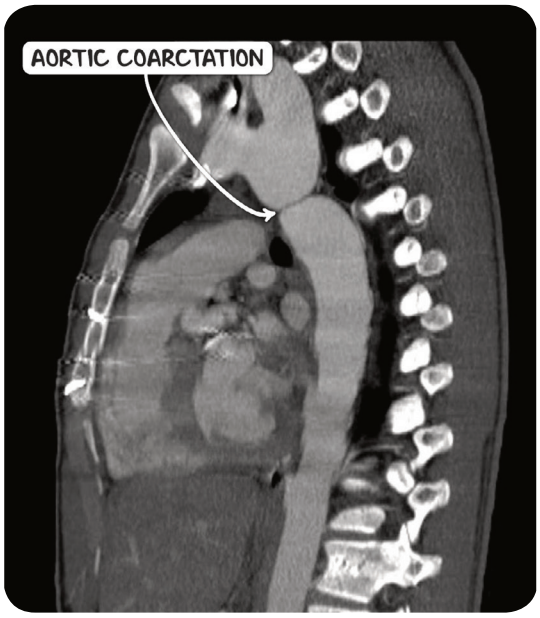

Figure 2.6 CT scan in the sagittal plane demonstrating coarctation of the aorta.

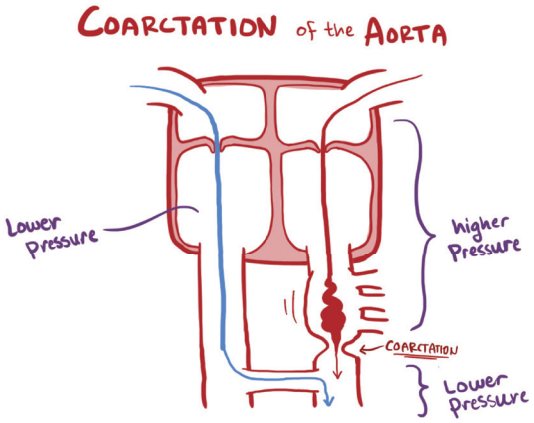

Figure 2.5 Illustration showing narrowing of aorta lumen.

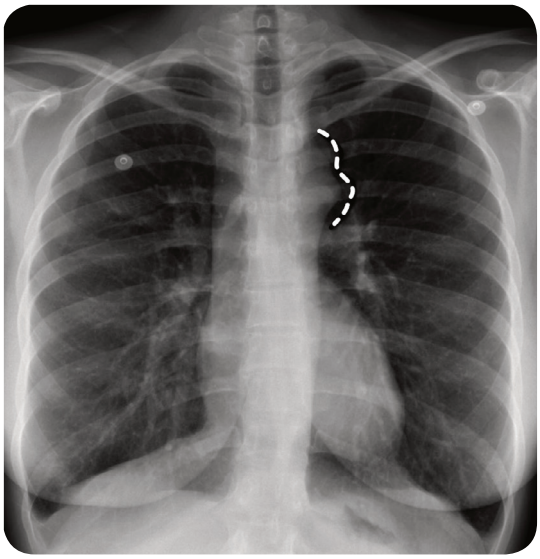

Figure 2.7 A chest radiograph demonstrating the figure of three sign seen in coarctation of the aorta.

PATENT DUCTUS ARTERIOSUS (PDA)

osms.it/patent-ductus-arteriosus

PATHOLOGY & CAUSES

- Ductus arteriosus remains open after birth
- Left-to-right shunt between atria
- Sometimes presents with congenital defects (congenital rubella syndrome)

CAUSES

Congenital rubella

- Mother-fetal transmission of rubella in first trimester → cytopathic damage to blood vessels, ischemia to organs
- Prematurity
- Perinatal distress, hypoxia

SIGNS & SYMPTOMS

Depend on size of PDA

- Smaller
  - Usually asymptomatic
  - *Neonates:* holosystolic "machine-line" murmur on auscultation
  - Infants, children, adults: continuous murmur
- Moderate
  - Exercise intolerance
  - Continuous murmur
  - Wide systemic pulse pressure
  - Displaced ventricular apex
- Larger
  - *Infants:* leads to heart failure
  - *Children:* shortness of breath, fatigability, Eisenmenger syndrome

DIAGNOSIS

DIAGNOSTIC IMAGING

Echocardiograph

- 2D suprasternal echocardiogram

Chest X-ray

- Normal/cardiomegaly

OTHER DIAGNOSTICS

ECG

- Left ventricular hypertrophy, left atrial enlargement

TREATMENT

- *Small asymptomatic PDA:* monitor

MEDICATIONS

Neonates (10–14 days)

- Close PDA using prostaglandin inhibitor

Symptomatic moderate/large PDA

- During heart failure
  - Digoxin, furosemide

SURGERY

Symptomatic moderate/large PDA

- Closure recommended for symptoms of left-to-right shunting, left-sided volume overload, reversible pulmonary arterial hypertension
  - *Children < 5kg/11lbs:* surgical ligation
  - *> 5kg/11lbs (including adolescents/adults):* percutaneous occlusion, surgical ligation for large PDA

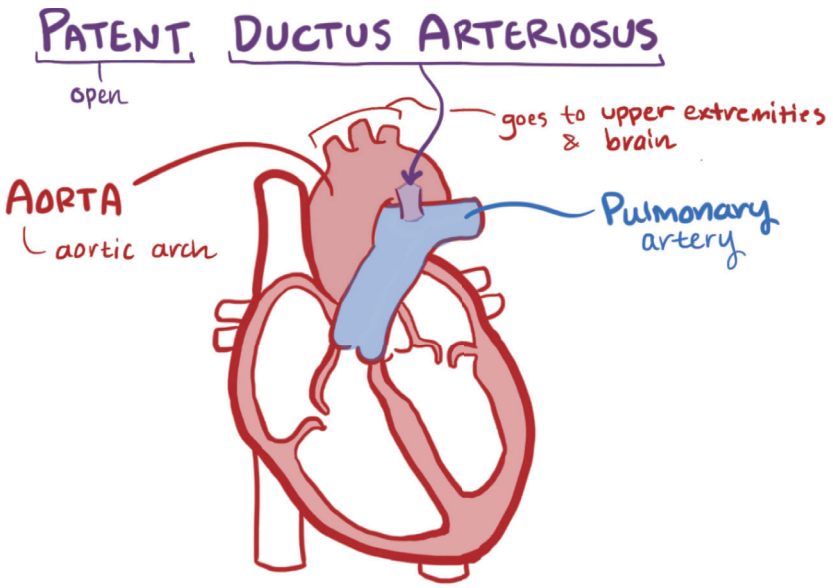

Figure 2.8 Illustration depicting location of a patent ductus arteriosus.

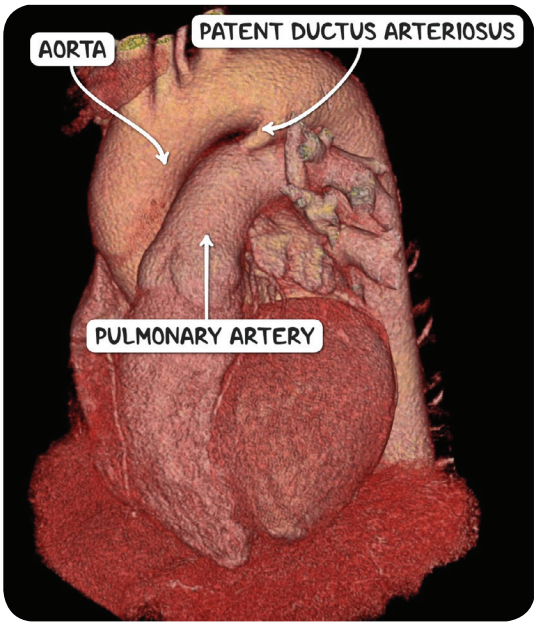

Figure 2.9 Volume-rendered CT scan of the heart and great vessels showing a patent ductus arteriosus.

VENTRICULAR SEPTAL DEFECT (VSD)

osms.it/ventricular-septal-defect

PATHOLOGY & CAUSES

- Left-to-right shunt between ventricles
- Most common congenital heart disease
- Left-to-right shunt between ventricles
- Often presents with other defects (e.g. tetralogy of Fallot)

Size of defect

- *Small:* restrictive
  - Normal pressure maintained between ventricles
- *Moderate or large:* non-restrictive
  - No pressure difference between ventricles

SIGNS & SYMPTOMS

- Asymptomatic in utero
- Holosystolic murmur (loud, high-pitched) located at lower left sternal border

Size of defect

- *Small:* asymptomatic, murmur
- *Moderate–large:* sweating, poor feeding/failure to thrive, respiratory infections. Murmur plus thrill, and diastolic rumble in mitral area
  - Signs of congestive heart failure (dyspnea, persistent cough, pulmonary vascular resistance)
  - Eisenmenger's syndrome

DIAGNOSIS

DIAGNOSTIC IMAGING

Chest X-ray

- Unreliable; may indicate left atrial enlargement, right ventricular hypertrophy, left ventricular hypertrophy, or pulmonary artery enlargement

Echocardiogram

- Determines location and size

MRI

- Use if echo does not diagnose

SURGERY

Cardiac catheterization

- Used if echo and MRI did not diagnose, but individual still has pulmonary hypertension

OTHER INTERVENTIONS

ECG

- Left ventricular hypertrophy
  - May see right ventricular hypertrophy; left, right atrial enlargement (may see Katz–Wachtel phenomenon)

TREATMENT

- Most small VSDs close on their own

SURGERY

- Repair larger shunts by age 2 to prevent pulmonary hypertension

Surgical repair

- Patch closure over ventricular septal defect (preferred treatment)

Transcatheter closure

- Mesh to close VSD (higher risk)

Hybrid procedure

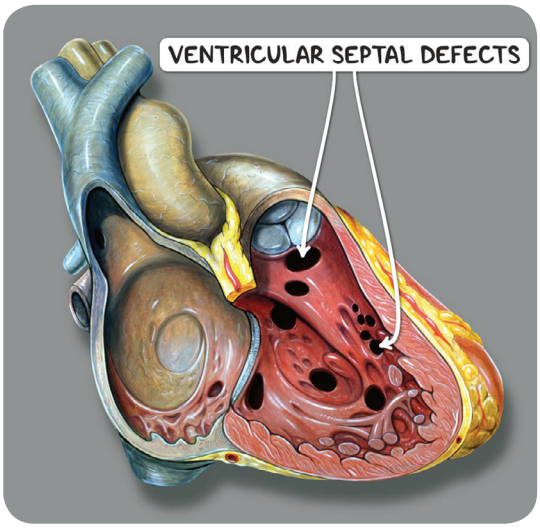

Figure 2.10 View of the right side of the heart showing multiple ventricular septal defects.

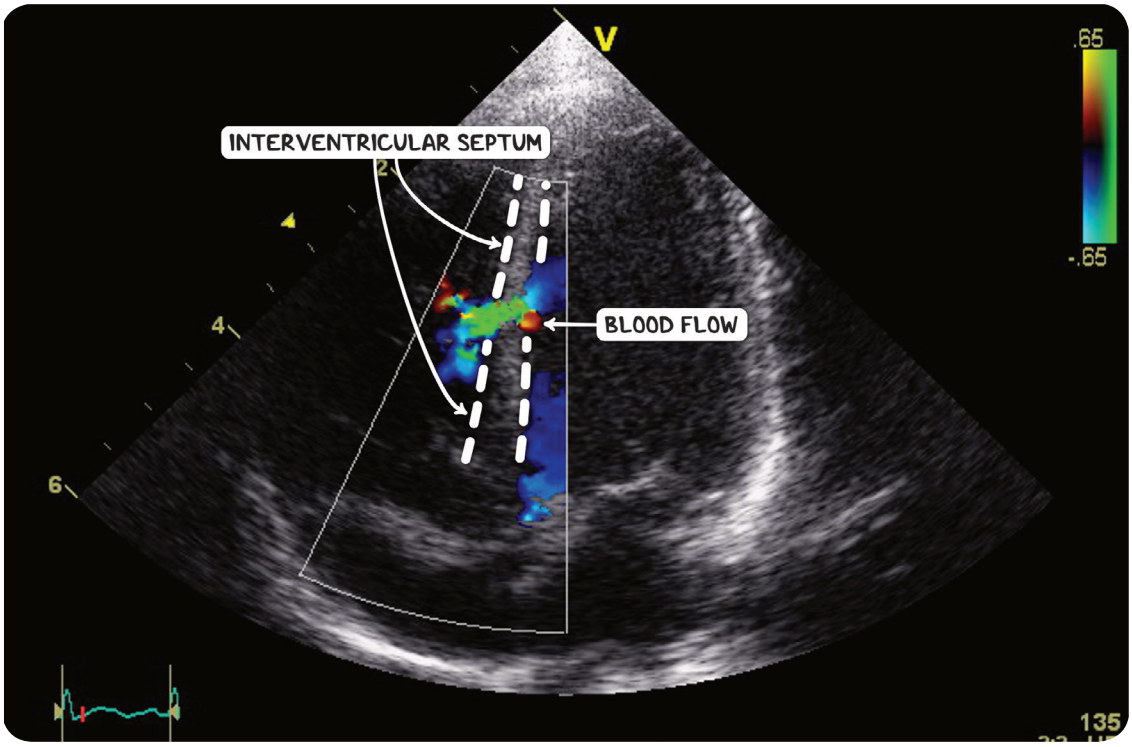

Figure 2.11 Doppler ultrasound scan demonstrating flow of blood across the interventricular septum in a VSD.