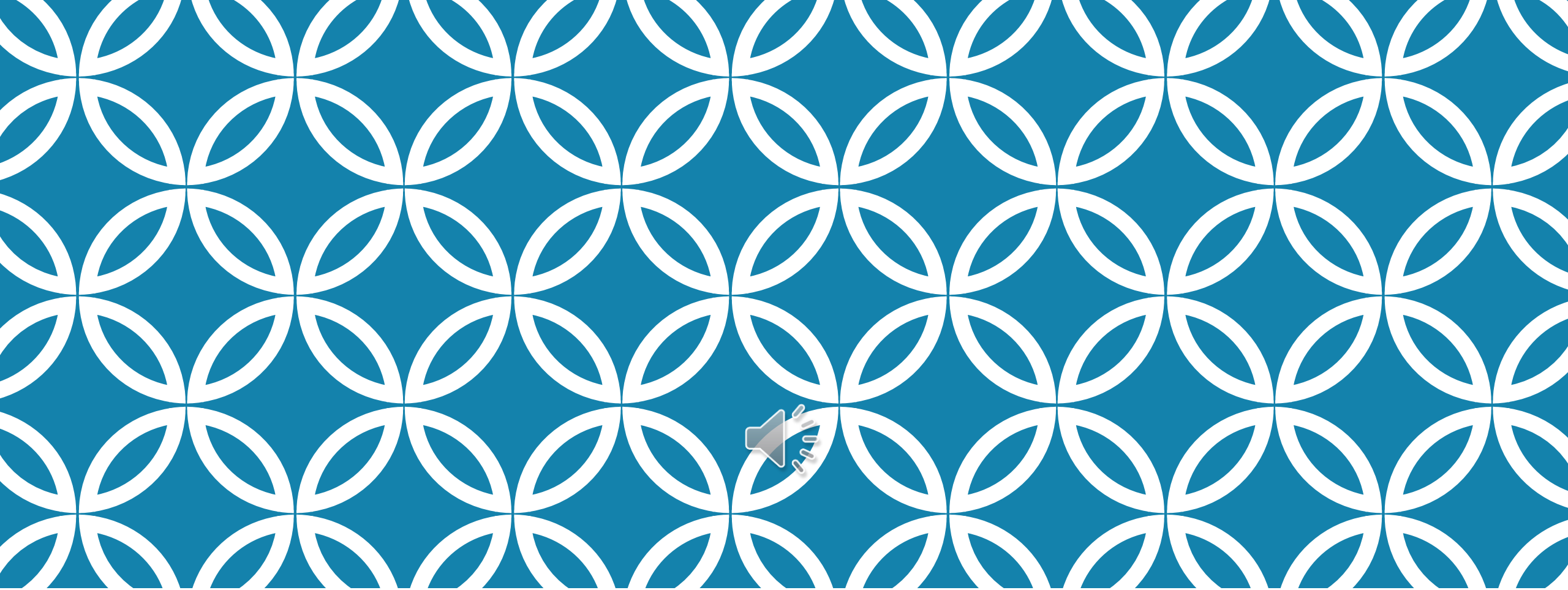


CARDIAC SURGERY

Dr. Bassam M. Hassan

PRESENTATION AND INITIAL MANAGEMENT

ASDs rarely cause symptoms in children, and are usually found incidentally. presenting in their third/fourth decade with fatiguability and reduced exercise tolerance. Rarely, if the defect is very big, there may be heart failure and failure to thrive (1% of patients). In older children and adolescents, there may be reduced exercise tolerance in comparison to peers. Patients are pink, with normal respiratory rate. (Infants may be tachypneic.) If desaturated, indicates right-to-left shunt, and should be further investigated before surgery.

Systolic ejection murmur over left second intercostal space—pulmonary flow murmur.

INVESTIGATIONS

- EKG normal in 20–40% of children. May show right axis deviation
- CXR shows iRA, RV, PA, and pulmonary vascular markings.
- Echo is diagnostic and identifies RA, RV, and PA enlargement, septal defect and associated anomalies.
- Catheterization is not indicated in the routine management of secundum ASD. It is necessary if there are concerns about pulmonary hypertension, to measure pulmonary vascular resistance and its reversibility with vasodilators.

TREATMENT

Percutaneous options

Defects up to 40mm within the fossa ovalis, with a 5mm rim between the defect and AV valves/major veins, can be closed percutaneously in the cath lab. The device is placed across the atrial septum via a catheter in the femoral vein, under TEE guidance. Success depends on the anatomy of the defect, but this is becoming the standard treatment for suitable defects.

Complications: **incomplete defect closure, device migration or erosion, local vascular complications.**

Surgical management

Up to 40% of small and moderate sized ASDs close spontaneously by age 4, although this becomes rare after age 2. Secundum ASDs still present after this time should be closed.

Indications

• In adults ASDs should be closed to prevent pulmonary hypertension.

• In older patients with pulmonary hypertension, if PVR <10U/m2, the outcomes are good; but with 15U/m2 or higher, the mortality is high.

• In infants, a large shunt with failure to thrive, recurrent respiratory tract infections or signs of heart failure. Evidence of RV or LV volume

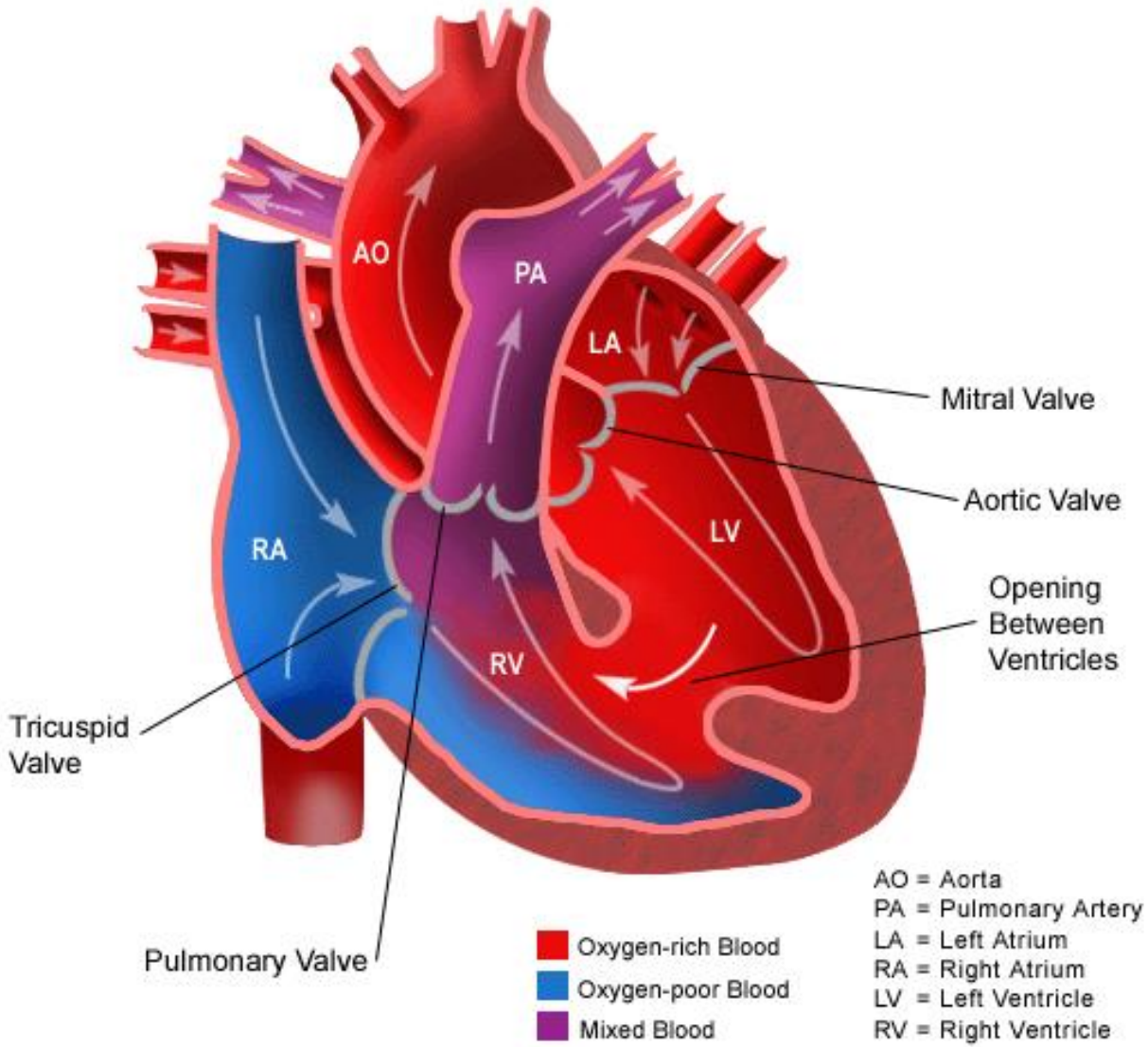
overload. Secundum ASDs should be closed electively in 3–5-year-olds.

• Paradoxical embolus causing TIA/stroke.

VENTRICULAR SEPTAL DEFECTS

- A VSD is a congenital, abnormal defect in the ventricular septum allowing communication of blood between the ventricles.
- They occur as isolated lesions, or in combination with other anomalies constituting 25% of all defects: the commonest congenital heart defect.
- Prevalence increasing over the last three decades with the improvement of imaging technology, and associated with maternal drug and alcohol abuse.

Ventricular Septal Defect (VSD)



VSDs CAN MOST SIMPLY BE CLASSIFIED AS FOLLOWS:

Perimembranous (70–80%)

Perimembranous VSDs: where a margin of the VSD consists of fi brous continuity between the tricuspid and aortic valves. The conduction bundles

run along the inferior rim of the defect. They may be *inlet, outlet, or inlet–outlet VSDs*.

Muscular (5–10%)

• Also called *trabecular* and including central, mid-muscular, apical, marginal, and multiple or 'Swiss cheese' VSDs. • They may be single or multiple, associated with other types of VSD and may occur anywhere within the muscular septum.

• The rim of the VSD is entirely muscular. The conduction bundles are remote from the defect, and in the case of the inlet VSD, they run near the superior margin of the defect. They may be located in the

inlet, apicotrabecular, or outlet portions of the RV.

Juxta-arterial (5–10%)

• Also called *conal septal, supracristal, infundibular, subpulmonary, doubly committed sub-arterial (DCSA) VSDs*.

• The conjoined leaflets of aortic and pulmonary valves form rim of VSD.

• The conduction bundles are remote from the defect.

• They extend up to the aortic and pulmonary annuli, and result in aorticvalve prolapse in up to 50% of cases.

PATHOPHYSIOLOGY

May be *restrictive defect*: small defects, presenting resistance to flow

across the defect, or *non-restrictive defect*: where the cross-sectional area of the defect is equivalent or larger than that of the aortic annulus

Three hemodynamic consequences:

(1) LV volume overload,

(2) increased pulmonary blood flow,

(3) reduced systemic cardiac output.

Presentation

Larger defects present in infancy with CHF, recurrent chest infections, failure to thrive; Qp;Qs >2 is poorly tolerated. Smaller defects may be entirely asymptomatic. Bacterial endocarditis is a risk.

Examination shows failure to thrive (FTT), tachypnea, and recession.

Investigations

- EKG shows LV and RV hypertrophy.
- CXR may show increased vascular markings.
- Echo is diagnostic, and defines the location and size of the defect, the extent of the hemodynamic consequences, and associated anomalies.
- In cases where pulmonary vascular disease is suspected, or there are multiple VSDs, cardiac catheterization may be required.

Natural history

- 30–40% of all VSDs close spontaneously. Up to 70% of small VSDs close spontaneously (50% of small PM VSDs close spontaneously in the first 2 years). Inlet perimembranous VSDs and DCSA VSDs do not close spontaneously, and are candidates for early closure.
- 10% of infants with large untreated VSDs die within fi rst year of life.
- Eisenmenger syndrome may be complicated by fatal hemoptysis, polycythemia, cerebral abscess or infarction, and RV failure.

TREATMENT:

Percutaneous closure

VSDs can be closed by percutaneously placed devices—these are mainly used to close muscular defects, and smaller perimembranous defects. In the latter there is a high incidence of AV node block, up to 10%.

Surgical closure

Operative mortality for VSD closure in pediatric setting is <1%.

Indications

- Moderate to large VSD after age 1 year.
- Congestive cardiac failure resistant to medical therapy in infants.
- Elevated pulmonary vascular resistance after the age of 6 months.
- Evolving aortic regurgitation in outlet VSDs.
- Multiple muscular VSDs (Swiss cheese defects) with significant shunt

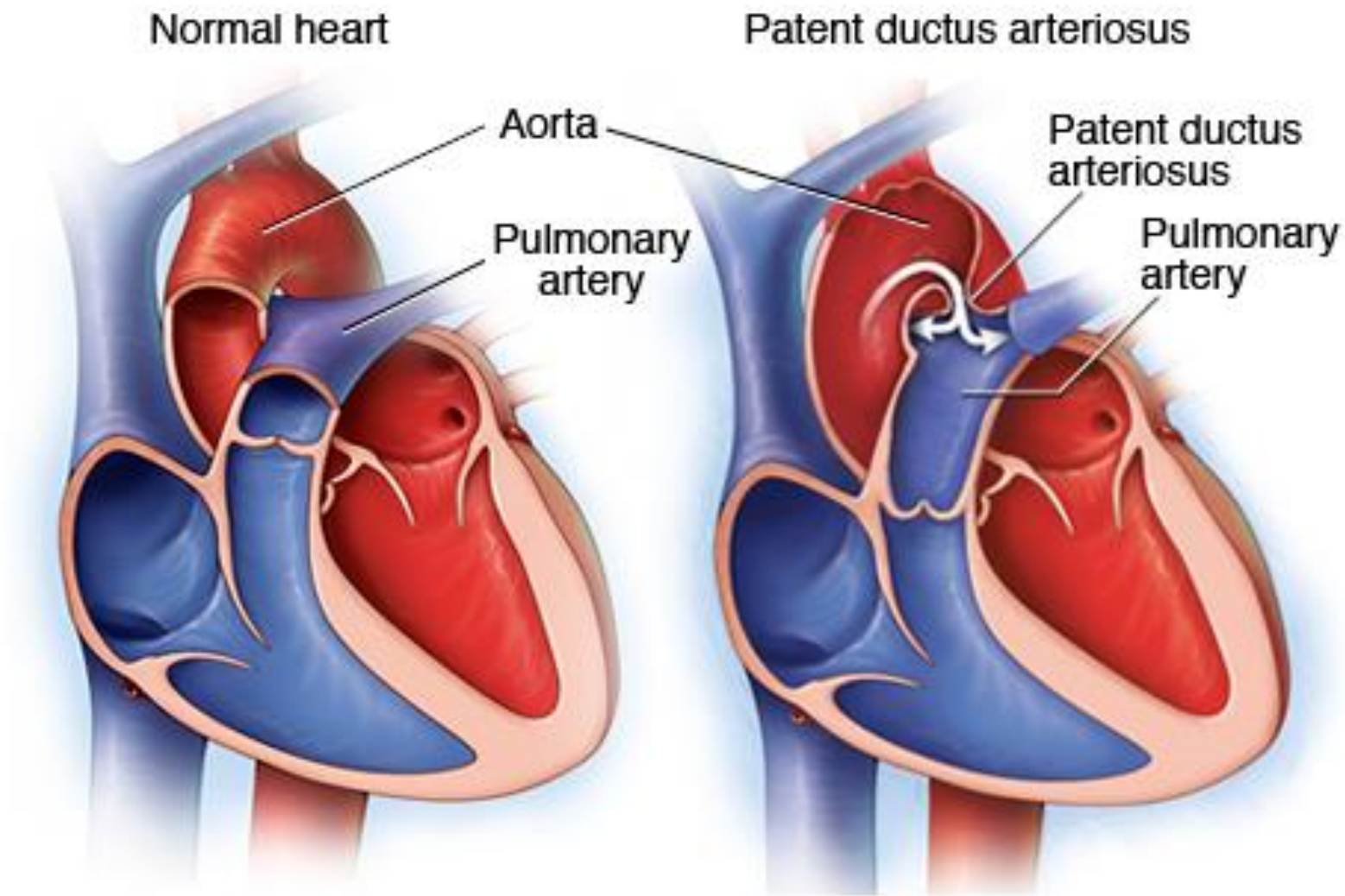
undergo PA band

PATENT DUCTUS ARTERIOSUS

• The arterial duct connects the pulmonary artery with the descending aorta, and in fetal life shunts blood from the pulmonary artery to the aorta, bypassing the immature fetal lungs.

• Most close within 2–3 weeks of birth, initially by contraction of the medial muscle of the duct, then by intimal hyperplasia. Persistence of the lumen after this time, with reversal of the flow, is called *patent ductus arteriosus (PDA)*.

• This anomaly makes up 12–15% of congenital heart defects, and up to 30% of defects in premature infants. 80% of premature infants <1200g present with this. Isolated PDA occurs 1 in 2000 live births.



PATHOPHYSIOLOGY

• When the PDA is large, aortic and PA pressures are equal, and the magnitude and direction of shunting depends on the PVR.

• As neonatal PVR falls the left-to-right shunt increases and congestive cardiac failure ensues.

• Pulmonary vascular changes may lead to irreversible pulmonary hypertension and reversal of the left-to-right shunt (Eisenmenger syndrome) which has a very poor prognosis, even though the initial reduction in the left-to-right shunt leads to a brief improvement in the clinical picture

Presentation

Presentation depends on the size of the PDA.

- **Large PDAs present with symptoms of severe heart failure in preterm babies, a wide pulse pressure, increase JVP, and a systolic murmur.**
- **The findings are similar in smaller PDAs but develop later; additionally the ductus may be calcified.**
- **Premature infants with this defect may be ventilator-dependent, or require long periods of non-invasive ventilation.**
- **Many children may be asymptomatic. Large shunts may present as congestive heart failure early in life.**

Examination

- **Bounding pulses due to wide pulse pressure.**
- **In premature infants, there is usually a systolic murmur at the second left intercostal space; with increasing size of the shunt, this murmur may extend into diastole and become the classic harsh, continuous 'machinery' murmur.**

Investigations

- The EKG shows left and sometimes right ventricular hypertrophy.
- CXR may be normal, or show cardiomegaly, an enlarged PA, and pulmonary congestion in large shunts.
- Echo is diagnostic, demonstrating the shunt flow from descending aorta to pulmonary artery, and enlarged left-sided chambers. Associated anomalies must be carefully excluded.
- Cardiac catheterization is not usually required, but catheter closure is usually possible in infants >2kg.

MANAGEMENT

Indomethacin and ibuprofen are equally effective in closing the patent duct in pre-term infants, and improve outcomes even in asymptomatic pre-term infants.

If medical therapy fails, surgical ligation is considered for all symptomatic infants, and in all infants in whom the duct has not closed after 3 months.

Indications for closure

- Persistent PDA is an indication for elective closure.
- Urgently if there is evidence of cardiac failure.

Atrioventricular septal defects

• AVSDs, also called *endocardial cushion defects, AV canal defects,* or *atrioventricular communis*, are a spectrum of defects resulting from incomplete development of the atrial septum, the inflow portion of the ventricular septum, and the atrioventricular valves.

• These defects make up about 5% of congenital cardiac anomalies.

• Congenital heart disease is present in 45% of children with Down's syndrome, and 45% of these have an AVSD.

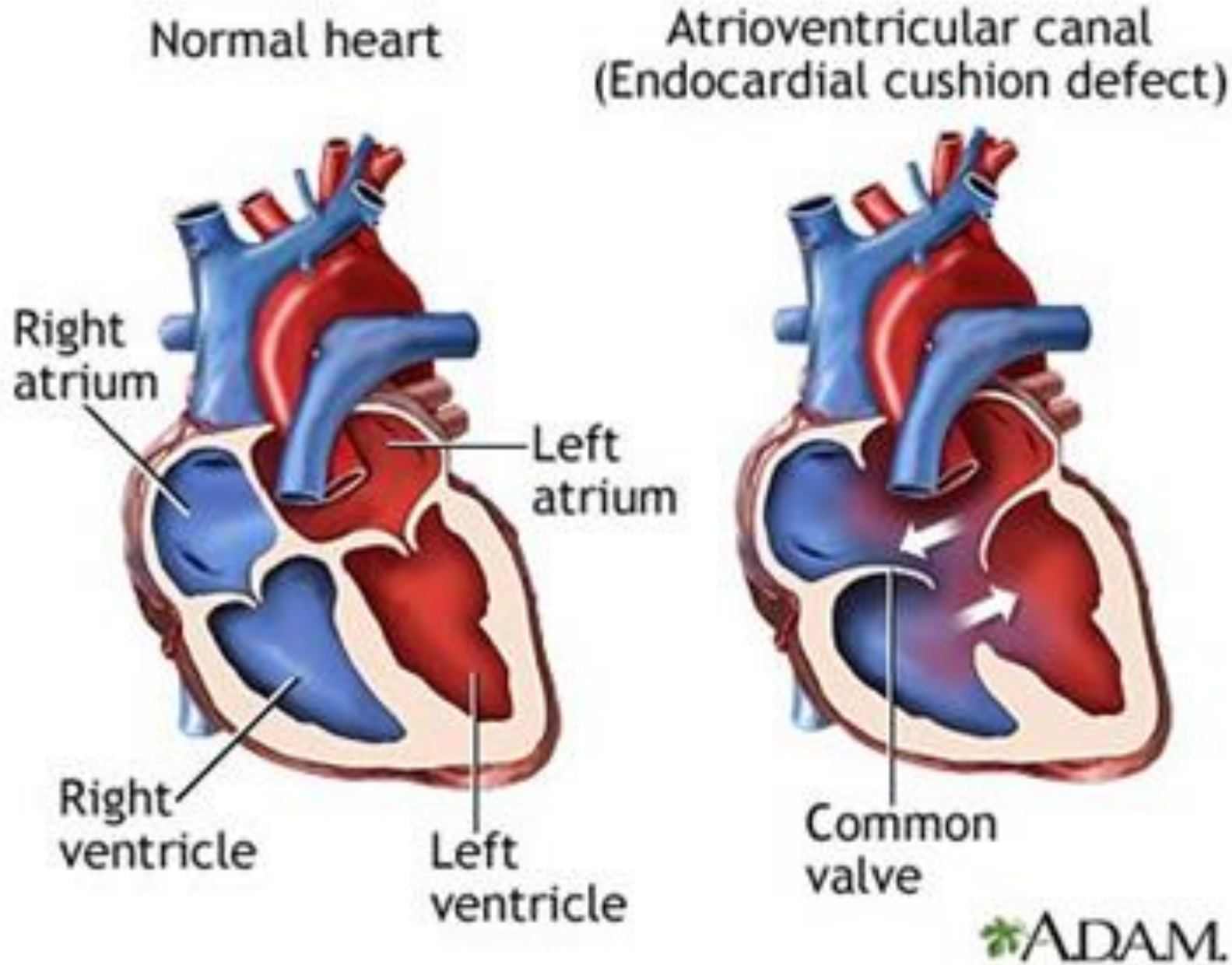
TYPES

Partial AVSD (primum ASD)

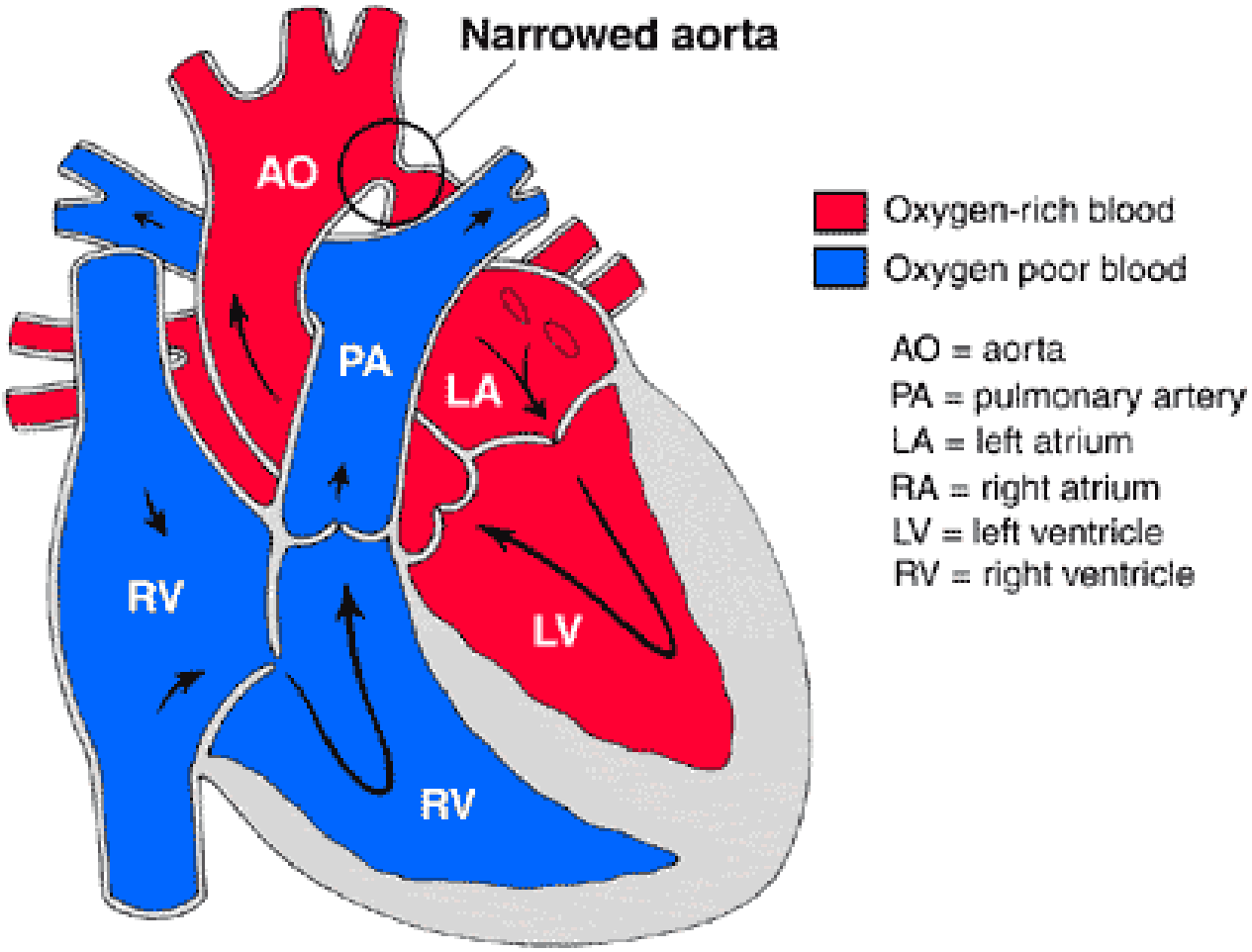
Inferior ASD immediately superior to the separate left and right AV valves; the left AV valve is trileafl et, and in 10% it is incompetent.

Complete AVSD

Single AV valve common to the right and left atrioventricular junction


Normal heart
Atrioventricular canal
(Endocardial cushion defect)
Right
atrium
Left
atrium
Right
ventricle
Left
ventricle
Common
valve
A.D.A.M.

Coarctation of Aorta



COARCTATION OF THE AORTA

• Coarctation of the aorta is defined as a congenital narrowing of the upper descending aorta, opposite the ductus arteriosus.

• This accounts for 5–8% of all congenital heart defects.

• It may be isolated, but is associated with bicuspid aortic valve and VSD.

It is the most common cardiac defect in *Turner syndrome*. (15–20%),

PATHOPHYSIOLOGY

• The hemodynamic consequences are high afterload on the LV,

increasing LV wall stress and causing *LV hypertrophy*.

Hypertension develops due to the mechanical obstruction and

possibly, renin–angiotensin-mediated pathways.Systemic perfusion depends on ductal flow

PRESENTATION AND NATURAL HISTORY

- Depends on the existence of coexisting abnormalities, as well as the location and severity of the location:

• *Neonates*: collapse, acidosis, hypotension, heart failure; absent femoral pulses on routine review.

• *Infancy*: upper extremity hypertension with absent/reduced femoral pulses; congestive heart failure causing dyspnea and failure to thrive.

• *Children/young adults*: headaches, lower extremity weakness, exertional dyspnea, fatigue; hypertension.

- Examination:

findings also depend on the age and presentation:

- In the shocked neonate, all pulses may be weak; however, absent femoral pulses should not be disregarded.
- There may be differential cyanosis, Systolic murmur, discrepancies of >20mmHg between upper and lower

INVESTIGATION

- EKG: LV hypertrophy.
- CXR: in sick children, cardiomegaly, pulmonary congestion.
- Echo: shows the arch,
- MRI/CT useful in older patients or reoperations to plan approach.

MANAGEMENT

Neonates

In the shocked neonate, the initial management is supportive, improving peripheral perfusion by *reopening the duct* if possible

Older children

Balloon angioplasty of the coarctation is now the fi rst-line treatment.

Hypertension should be treated, usually with β-blockers, but intervention should not be delayed while waiting for normotension.

Indications for surgery

Isolated coarctation is an indication for surgical repair, once critically ill infants have been stabilized, and within 4–6 weeks of presentation in stable children.