Acyanotic congenital heart disease

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- Atrial septal defect (ASD)
- Atrioventricular canal
- Ventricular septal defect (VSD)
- Patent ductus arteriosus (PDA)
- Congenital aortic stenosis
- Congenital pulmonary stenosis
- Coarctation of aorta
- Congenitally corrected transposition of great arteries (L-TGA)
- Congenital anomalies of coronary artery

Atrial septal defect (ASD)

- **4 types : *1) secundum* (most common, located at middle portion of atrial septum involving fossa ovalis)**
  2) *primum* (located at inferior atrial septum)
  3) *sinus venosus* (located adjacent to superior vena cava, usually associated with anomalous pulmonary venous drainage)
  4) *coronary sinus*

- ***Lutembacher syndrome* = ASD + mitral stenosis (ASD ameliorates mitral stenosis and mitral stenosis augments left-to-right interatrial shunt)**

- ***Symptoms:*** majority female. Second heart sound widely split and fixed.
- Children and adolescent: usually asymptomatic
- Adult: palpitation, exertional dyspnea, fatigue, recurrent lower respiratory infection, heart failure (due to atrial flutter/fibrillation), coronary artery disease, hypertension, paradoxical embolism
- Primum ASD → mitral regurgitation
- Secundum and sinus venosus ASD → left-to-right shunting → right ventricular overload, enlargement of right atrium and ventricle, pulmonary artery, pulmonary hypertension → pulmonary vascular obstructive disease

- ***Management:***
- **Patent foramen ovale (= extremely small ASD) need observation**
- **All hemodynamically significant ASD (Qp/Qs or pulmonary to systemic flow >2) should be closed by percutaneous device or surgical repair. If present, anomalous pulmonary veins are incorporated into left atrium. Mitral regurgitation may require repair.**
- **Patients with right-to-left shunting are inoperable (because right-to-left shunt maintain cardiac output at expense of desaturation) → heart-lung transplantation**
- **If no associated valve abnormalities, antibiotic prophylaxis for subacute bacterial endocarditis (SBE) not recommended for secundum and sinus venosus ASD 6 months after surgery. Primum ASD need continue prophylaxis because of left atrial ventrciular valve abnormality.**

Atrioventricular septal defect = atrioventricular canal

= more extensive primum ASD + atrial ventricular (AV)valve abnormalies + inlet VSD

- **Majority has Down syndrome**
- **Classified as complete, partial and intermediate types**
- **Symptoms: growth failure, frequent respiratory infection**
- **Left-to-right shunt, AV valve insufficiency → pulmonary hypertension → pulmonary vascular obstructive disease, heart failure**
- **Treatment: surgical repair (rarely reach 4th decade without surgery). Follow-up include: AV valve insufficiency (or rarely stenosis), subaortic obstruction, atrial or ventricular arrhythmias and may need reintervention.**

Ventricular septal defect (VSD)

= most common congenital heart disease at birth

- **Classified as: perimembranous (80%, defect surrounding membranous septum), supracristal, muscular and inlet (endocardial cusion) types**
- **Spontaneous closure (more common if small VSD) occur by age 3 (45%), occasionally until age 8-10 or even later. In adult is uncommon (<10%)**
- **Symptoms: growth failure, recurrent respiratory infectin, heart failure**
- **Holosystolic murmur at left lower sternal border (small VSD louder than larger ones).**
- **Small VSD (<0.5 cm^2) → small left-to-right shunting (Qp/Qs <2) → asymptomatic**

  moderate VSD (0.5-1 cm^2) → large left-to-right shunting (Qp/Qs >2) → pulmonary hypertension, mild heart failure

large VSD (>1 cm^2) → severe pulmonary hypertension, severe heart failure → pulmonary vascular obsturctive disease → bidirectional shunting → right-to-left shunting (=Eisenmenger's syndrome with cyanosis, polycythemia, dyspnea, exercise intolerance, hemoptysis, chest pain, brain abscess, endocarditis, arrhtyhmias, sudden death etc)

- Treatment: If Qp/Qs > 1.8 → surgical VSD closure
- Eisenmenger's syndrome → heart-lung transplantation (because VSD closure eliminates portion of cardiac output that is dependent on right-to-left shunting, so contraindicated). If less symptomatic → pulmonary vasodilator therapy
- Successful VSD repair → no more antibiotic prophylaxis

Patent ductus arteriosus (PDA)

- Most common congenital heart disease in neonates

= persistent communication between left pulmonary artery and descending aorta

- In fetus, patency of ductus arteriosus (evolves from 6th aortic arch connecting pulmonary artery to aorta during fetal life) maintained by arterial low oxygen saturation and elevated endogenously produced prostaglandins. At birth, prostaglandin fall and oxygen saturation rise → ductus close. In premature, prostaglandin continue high → patent ductus arterosus

- Symptoms : predominately female. Premature birth, maternal rubella, or birth at high altitude.
- Continuous machinery murmur at left upper sternal border
- Growth failure, fatigue, dissecting or rupture aneurysm of ductus, infective endarteritis or endocarditis
- Left-to-right shunting → pulmonary hypertension, heart failure
- Treatment: premature neonates – indomethacin (80% may close)
- After infancy – catheter or surgical closure
- Very small PDA (silent ductus) – observation and antibiotic prophylaxis

Congenital aortic stenosis (AS)

(I) Valvular Aortic Stenosis :-

- **Systolic murmur at right and left upper sternal border**
- **Usually asymptomatic**
- **Symptoms:**

  exertional dyspnea **(due to increase left atrial pressure because of left ventricular pressure overload and hypertrophy, heart failure)**

  angina **(due to inadequate subendocardial blood flow)**

  syncope **(due to low cardiac output and ventricular tachyarrhythmias)**

  sudden death **(due to ventricular fibrillation)**
- ***Severity of AS*** **:**

  mild AS (left ventricular-aorta gradient < 25 mmHg)

  moderate AS (aortic gradient 25-50 mmHg)

moderately severe AS (aortic gradient 50-75 mmHg)

severe AS (aortic gradient > 75 mmHg)

- ***Management of AS :***

  mild AS – observation and antibiotic prophylaxis

  moderate to severe AS (aortic gradient > 50 mmHg)-

  symptomatic and need balloon valvuloplasty or surgical repair (valvotomy or replacement)

(II) Subvalvar aortic stenosis :-

= obstructive fibrous ridge immediately below aortic valve

(III) Supravalvar aortic stenosis :-

- very rare, usually associated with Williams syndrome (mental retardation, dental abnormality, elfin facies, small statue)
- in adult, associated with other stenoses as coronary, renal, mesenteric vessels etc.

Treatment: surgical repair

Congenital pulmonary stenosis (PS)

- **Classified as valvular, supravalvar and subpulmonary**
- **+ VSD = tetralogy of Fallot**
- **Mild and moderate PS usually asymptomatic. May deteriorate rapidly with dyspnea, fatigue, right ventricular failure, syncope, chest pain of right ventricular myocardial ischemia, sudden death.**
- **Systolic murmur at left upper sternal border**
- ***Severity of PS:***

  mild PS (right ventricle-pulmonary artery RV-PA gradient < 50 mmHg) – observation and antibiotic prophylaxis

  moderate PS (RV-PA gradient 50-70mmHg) – balloon valvuloplasty

  severe PS (RV-PA gradient > 70mmHg) – balloon valvuloplasty or surgical valvotomy or replacement

Coarctation of aorta

= narrowing of a segment of aorta at junction of ductus arteriosus and aortic arch

- Often associated with bicuspid aortic valve, VSD, AS, mitral valve abnormality
- Symptoms: infant – severe heart failure and very poor prognosis without surgery

  child/adult – headache, epistaxis, leg fatigue, hypertension in upper extremity but decreased femoral and pedal pulses, heart failure, coronary artery disease, stroke, differential cyanosis (upper body pink and lower body blue), rupture or dissection of paracoarctation aorta, infective endarteritis or endocarditis, cerebral hemorrhage due to rupture of cerebral aneurysm
- Treatment: surgical repair (untreated coarctation mean survival = 35 years, with 75% mortality by 50 years), balloon angioplasty or stenting.

Congenitally corrected transposition of great arteries (= L-transposition of great arteries)

- **Uncommon**
- **Characterized by discordance between atria and ventricles, as well as between ventricles and great arteries. Therefore, desaturated blood from right atrium → right-sided mitral valve → morphologic left ventricle →pulmonary artery → pulmonary vein → left atrium → left-sided tricuspid valve → morphologic right ventricle → aorta. Physiologic blood flow remains normal, corrected by the transposed great arteries.**
- **Often associated with VSD, PS, left AV regurgitation, heart block**
- **Symptoms: left AV valve regurgitation, heart block, failure of right (systemic) ventricle, heart failure**
- **Treatment: valve repair or replacement, double switch operation, heart transplantation**

Congenital anomalies of coronaries

(1) ***Abnormal origin of coronaries from aortic sinuses:***

a) **Origin of left main coronary from right coronary sinus:**

→ **ischemia, sudden death**

b) **Origin of right coronary from left coronary sinus or left main coronary artery:**

→ **ischemia, sudden death**

(2) Abnormal origin of coronaries from pulmonary artery:

- **Uncommon**
- **Ischemia, myocardial infarction, mitral regurgitation, heart failure, sudden death**

(3) Coronary artery fistula:

= communication between coronary artery and another portion of heart eg. right atrium (mostly), coronary sinus, right ventricle, pulmonary artery, left atrium, left ventricle etc.

Treatment = surgical closure of fistula

(4) ***Coronary artery abnormality associated with congenital heart disease:***

eg. Tetralogy of Fallot, pulmonary atresia, D-transposition of great arteries