The Heart

Congenital heart disease (CHD)

- A cause of heart failure in children
- Consequences depend on lesion:
  - Location
  - Size
  - Nature of abnormality

Causes of CHD

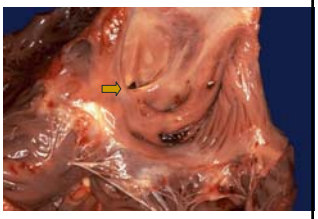
- 80% unknown
- Infection – rubella
- Chromosomal (Turners syndrome XO – coarctation of aorta, Downs trisomy 21 – ASD/VSD/PDA)
- Poorly defined familial susceptibility

Clinical effects of CHD

- Failure to thrive
- Cyanosis (R – L shunt)
- Cardiac failure
- Pulmonary hypertension
- Infective endocarditis

Atrial septal defect (ASD)

- Most common is a patent foramen ovale (overlap of septum primum/septum secundum)
- Late effect – pulmonary hypertension

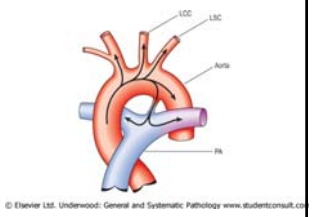


Ventricular septal defect (VSD)

- Always some L – R shunting
- Most occur in superior (membranous) septum and can have associated mitral valve abnormality

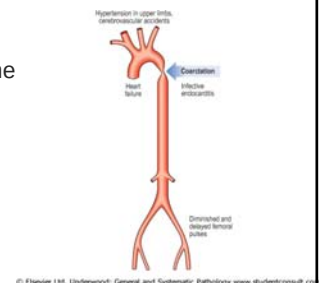
Patent ductus arteriosus

- L – R shunt
- Pulmonary hypertension



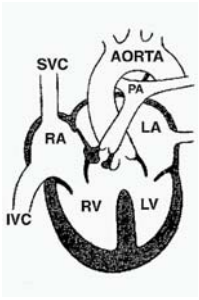
Coarctation of aorta

- Upper limb hypertension
- Turner's syndrome association



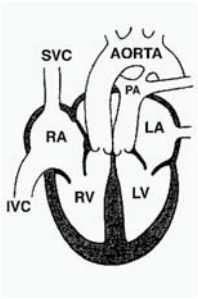
Tetralogy of Fallot

1. VSD
2. Overriding aorta
3. Pulmonary stenosis
4. Right ventricular hypertrophy



Transposition of great vessels

- Switching of aorta and pulmonary trunk
- Immediately lethal unless combined wih another defect – PDA, ASD,VSD

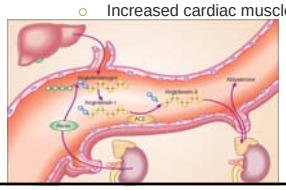


Cyanosis in CHD

- Circulating hypoxic blood
- Present in Fallot tetralogy & transposition
- Others (e.g. VSD) sometimes become cyanotic with reversal of flow through defect (right ventricular hypertrophy)

Cardiac failure

- End result of many pathological processes
- Leads to complex adaptive processes
  - Increased sympathetic tone
  - Antidiuretic hormone secretion
  - Increased renin-angiotensin activity
  - Increased cardiac muscle bulk



Right and left heart failure

- Interrelated but can be distinct esp. in early stages
- Left – pulmonary congestion/oedema
- Right – systemic congestion (↑ jugulovenous pressure), hepatomegaly
- “Congestive cardiac failure” (CCF) - both

Cardiac output

- Usually decreased in cardiac failure
- High output failure caused by:
  - Increased blood volume
  - Anaemia (severe)
  - Cirrhosis (vasodilatation with decreased peripheral resistance)

Causes of cardiac failure

- Hypertension
- Valve disease
- Lung disease
- Ischaemic heart disease
- Lung disease
- Cardiomyopathy

Pathological changes

- As for causative condition + ventricular hypertrophy/dilatation
- Pleural effusion

Ischaemic heart disease

- Coronary atherosclerosis
  - Acute infarcts
  - Fibrosis
    - Localised (healed infarct)
    - Diffuse (chronic ischaemia)

Commonest cause of death in Europe & N. America

Ischaemia

- Failure of blood supply to keep up with demand
  - Decreased supply – vessel narrowing (atherosclerosis)
  - Increased demand – muscle hypertrophy, as in hypertension

Coronary atherosclerosis

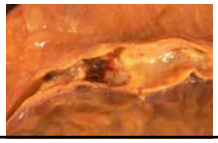
- Affects large proximal vessels
- 75% occlusion almost always gives significant clinical ischaemia. Cardiac muscle cells have high aerobic energy demands

Routes to ischaemia

- Slow progressive vessel narrowing
- Major plaque ulceration/rupture with thrombosis (most common route to acute infarct)
- Occlusion of coronary ostia (syphilis)
- Hypotension (shock) – subendocardial infarct

Acute myocardial infarct

- Necrosis – release of muscle enzymes (diagnostically useful)
- Inflammatory cell infiltration (neutrophils)
- Fibrous repair



Location of infarct….

- Depends on
  - Vessel involved (L – anterior, R – posterior/inferior)
  - Degree of collateral circulation, if any (coronaries are normally end arteries)

Complications of myocardial infarction

- Heart failure
- Arrhythmia
- Mitral incompetence
- Cardiac rupture
- Embolism
  - Direct from thrombus in heart
  - Pulmonary thromboembolus

Chronic ischaemic heart disease

- Cardiac failure
- Angina

Rheumatic fever

- Annual incidence:
  - Western Europe 1/100,000
  - Sub-Saharan Africa 5,700/100,000
- Incidence decreases with improving social circumstances (less crowding)
- Individual (HLA) susceptibility also important

Group A β–haemolytic streptococcus

- All cases associated with recent infection (e.g. pharyngitis, pyoderma)
- Some bacterial serotypes (M antigen) are more significant in causing rheumatic fever
- Antibody and cellular immune response cross-reacts with human connective tissue

Clinically

- Joints (arthritis)
- Heart (arrhythmias etc.)
- Skin (erythema marginatum)
- Central nervous system (chorea)
- Mainly 5-15 years (20% adult)

"Licks the joints but bites the heart"

Pancarditis…

- Pericarditis
- Myocarditis
- Endocarditis – responsible for chronic valvular damage

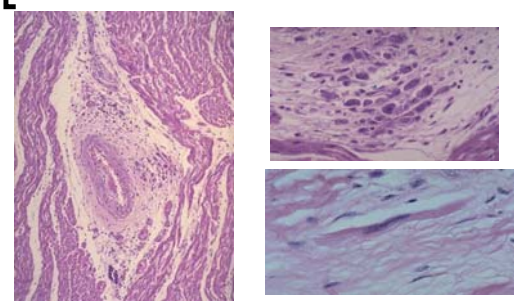
Acute, recurring, chronic

- Symptoms prone to recur with subsequent Strep. Infections
- Chronic disease leads to fibrosis (chordae of heart valves + valve cusps)

Histopathology

- Aschoff bodies (small granulomas around necrotic collagen – T cells, macrophages)
- Anitschkoff cell – an unusual spindly macrophage

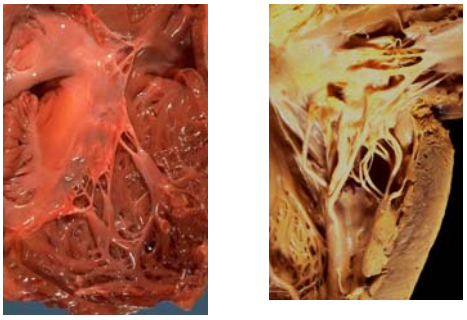
Aschoff nodule and Anitschkow cell



Rheumatic valve disease

- Most common lesion is mitral stenosis
- Aortic valve second most frequently involved

Normal vs. chronic rheumatic valve



Valvular heart disease

- 10% of heart failure caused by valve disease
- Abnormality may be congenital or acquired
- Acquired abnormality affects mainly the mitral and aortic valves

Valvular stenosis (acquired)

- Relatively few causes of stenosis
  - Postinflammatory (rheumatic) mitral or aortic stenosis
  - Calcific aortic stenosis (usually on congenitally bicuspid valve)

Valvular incompetence/regurgitation

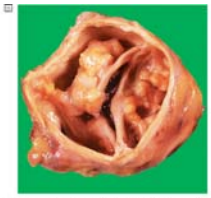
- Leaflet abnormality (rheumatic, infective endocarditis)
- Papillary muscle damage (ischaemia leading to mitral valve disease)
- Valve ring abnormalities (syphilitic aortitis, mitral ring dilatation with cardiac dilatation in congestive failure)

Mitral stenosis

- Rheumatic
- More common in females
- Leads to pulmonary hypertension, R ventricular hypertrophy

Aortic stenosis

- L ventricular hypertrophy
- Chest pain
- Sudden death

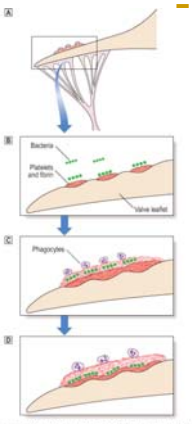


Infective endocarditis

- *Usually* involves a heart valve
- Risk is much higher with a diseased valve – infection occurs with non-virulent organisms (Strep. viridans)
- Normal valves can be infected in septicaemia with virulent bacteria (Staph. aureus)

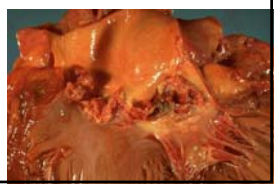
Pathogenesis

- Fibrin deposits on injured endothelium
- Circulating bacteria infect microthrombi
- Bacterial proliferation and inflammatory infiltration/tissue destruction



Complications

- Valvular incompetence
- Emboli
- Finger clubbing
- Glomerulonephritis



Non-infective cardiac vegetations

- Systemic lupus erythematosus
- Non-bacterial thrombotic endocarditis – seen in very ill people e.g. terminal cancer

Myocarditis

- Viral (influenza, ECHO, HIV, CMV)
- Trypanosomiasis (S. American, Chaga's disease – T cruzi)
- Non-infective (e.g. eosinophilic associated with parasites elsewhere, rheumatic, SLE)

Myocarditis (2)

- General effects of infection
- Arrhythmia
- Cardiac failure (acute or chronic)

Cardiomyopathies

- Myocardial diseases "of unknown cause" – excludes hypertensive, valvular, ischaemic

Cardiomyopathy

- Dilated (congestive)
  - Progressive congestive failure with a dilated heart (need to consider ischaemia, toxins, viruses)
  - Nutritional deficiency (protein, thiamine (B1), other vitamins)
  - Some cases apparently familial

Cardiomyopathy

- Hypertrophic cardiomyopathy
  - Asymmetric left ventricular hypertrophy
  - Affects septum
  - Associated with sudden death
  - Often familial with structural protein abnormalities (tropomyosin)

Restrictive cardiomyopathy

- A "stiff" heart with reduced filling in diastole
- Dilated atria
- Endomyocardial fibrosis (EMF,tropical)
  - Subendocardial fibrosis with thrombosis
- Loeffler endomyocarditis
  - Similar to EMF but with eosinophil infiltrate (possibly related to parasite infection)
- Amyloid heart disease has similar features

Pericarditis

- Acute – viral, pyogenic bacteria, TB
- Non-infective – rheumatic, SLE, after myocardial infarct
- Can heal by fibrosis – constrictive pericarditis