Heart disease: diseases of the heart structure, muscle and valves.

Christopher Whitty
Gresham College 2023

Three broad groups of things can lead to the heart failing.

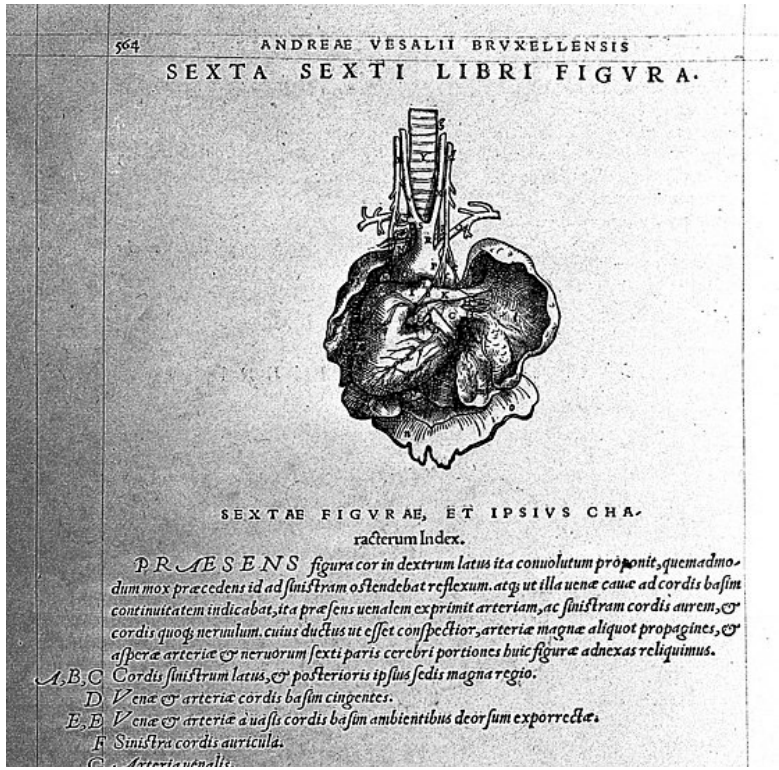
- Myocardia ischaemia- lack of oxygen usually due to narrow or blocked coronary arteries (coronary heart disease).

- The heart rhythm is abnormal- either too fast or too slow (arrhythmia).

- The structures of the heart are damaged including muscle and valves.

Leonardo da Vinci. Circa 1511-13.

The understanding of the structure and function of the heart changed around the time Gresham College was founded.
Andreas Vesaleus 1514–1564 and William Harvey 1578–1657.



564 ANDREAE VESALII BRVXELLENSIS

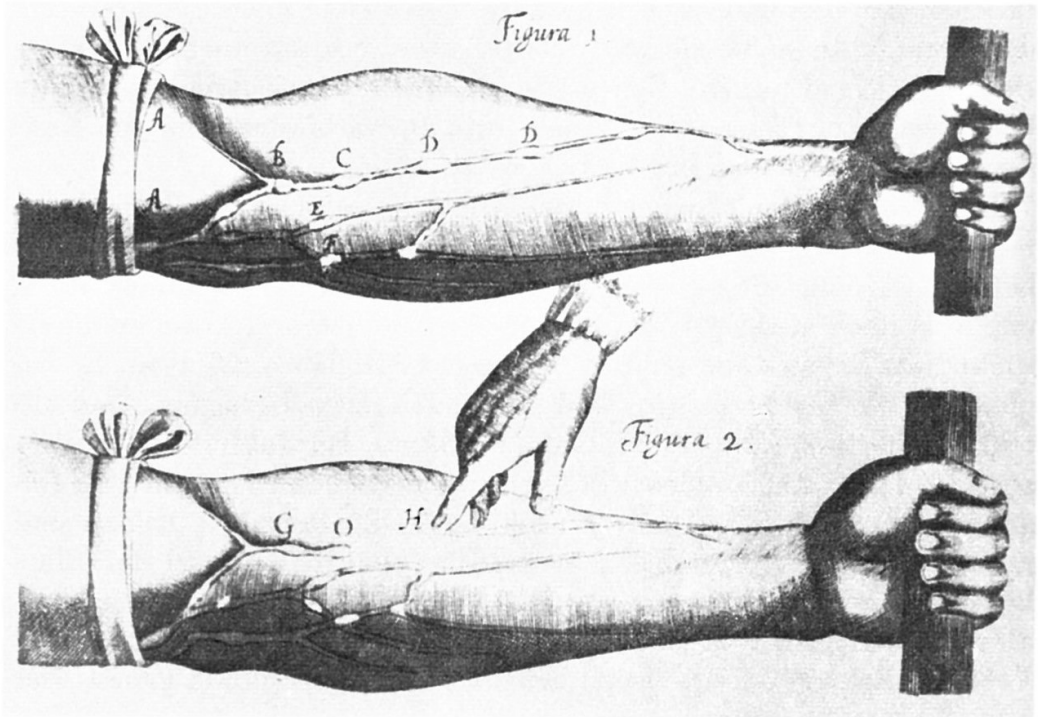
SEXTA SEXTI LIBRI FIGVRA.

SEXTAE FIGVRAE, ET IPSIVS CHA-
racterum Index.

PRAESENS figura cor in dextrum latus ita conuolutum proponit, quemadmodum mox præcedens id ad sinistram ostendebat reflexum. atq; ut illa uenæ cauæ ad cordis basim continuitatem indicabat, ita præsens uenalem exprimit arteriam, ac sinistram cordis aurem, & cordis quoq; nerulum. cuius ductus ut esset conspectior, arteriæ magnæ aliquot propagines, & asperæ arteriæ & neruorum sexti paris cerebri portiones huic figuræ adnexas reliquimus.

A,B,C Cordis sinistrum latus, & posterioris ipsius sedis magna regio.
D Venæ & arteriæ cordis basim cingentes.
E,E Venæ & arteriæ à uasis cordis basim ambientibus deorsum exporrectæ.
F Sinistra cordis auricula.

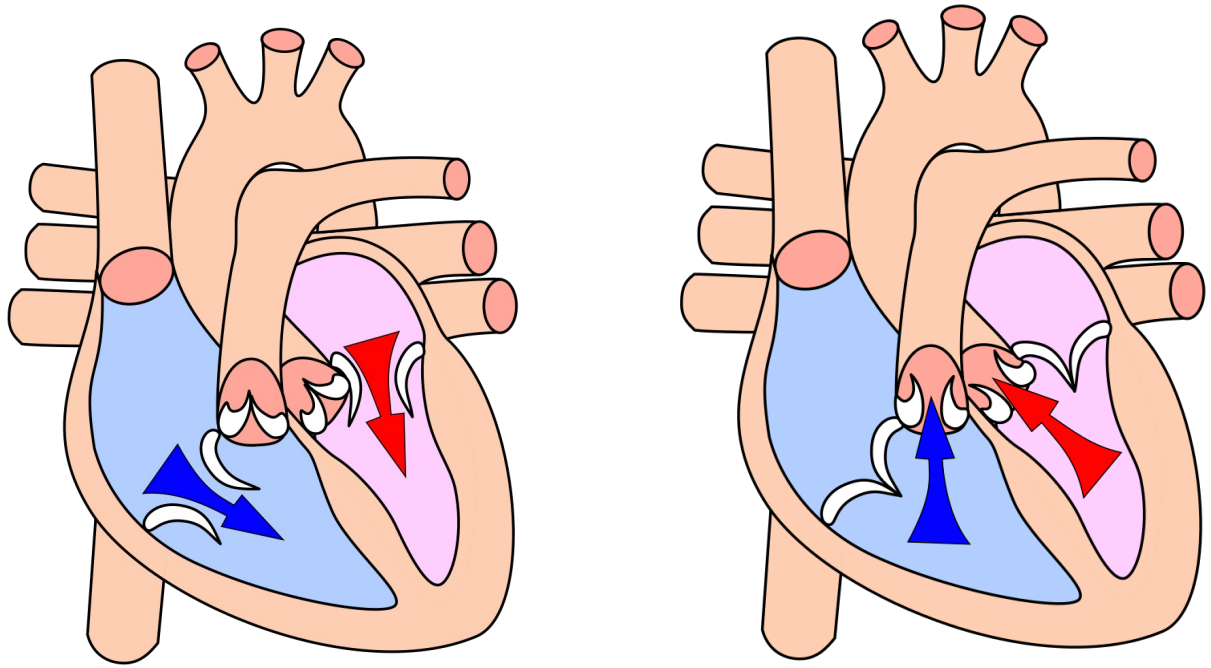
De Humani Corporis Fabrica,
Book 6. Andreas Vesalius 1543.



De Motu Cordis,
William Harvey 1628.

The structure of the heart as two pumps in one.

- Four chambers. Left and right sided systems.
- Upper pump (atrium) and lower pump (ventricle).
- A strong and relentless muscle usually contracting once a second or more.
- Between them are valves to ensure one-way flow.
- Around the heart is a covering- the fibrous pericardial sac.



Wapcaplet, Reytan, Mtcv

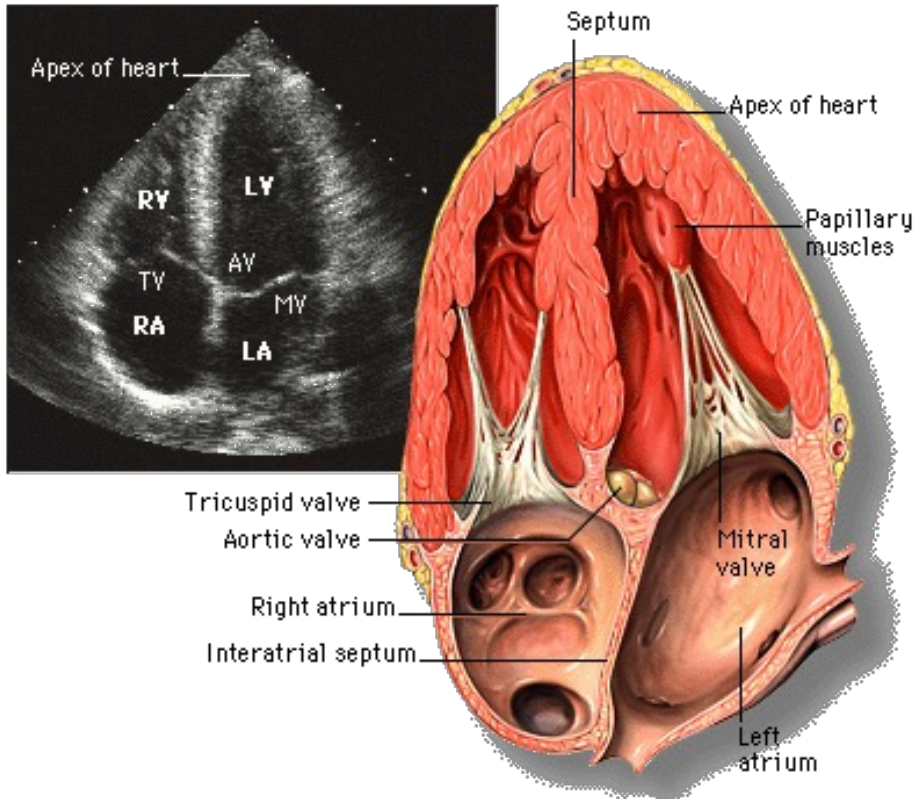
Diagnosing structural heart disease.

- Symptoms and timing- medical history.
- Physical examination. Heart size, pulse, blood pressure.
- Heart sounds for valvular disease.
- A major advance was the stethoscope.
- René Laennec in Paris in 1816, subsequently improved in stages from 1850s to 1980s.
- Plain chest X-ray for heart size and shape.
- ECG.

Science museum / US gvt

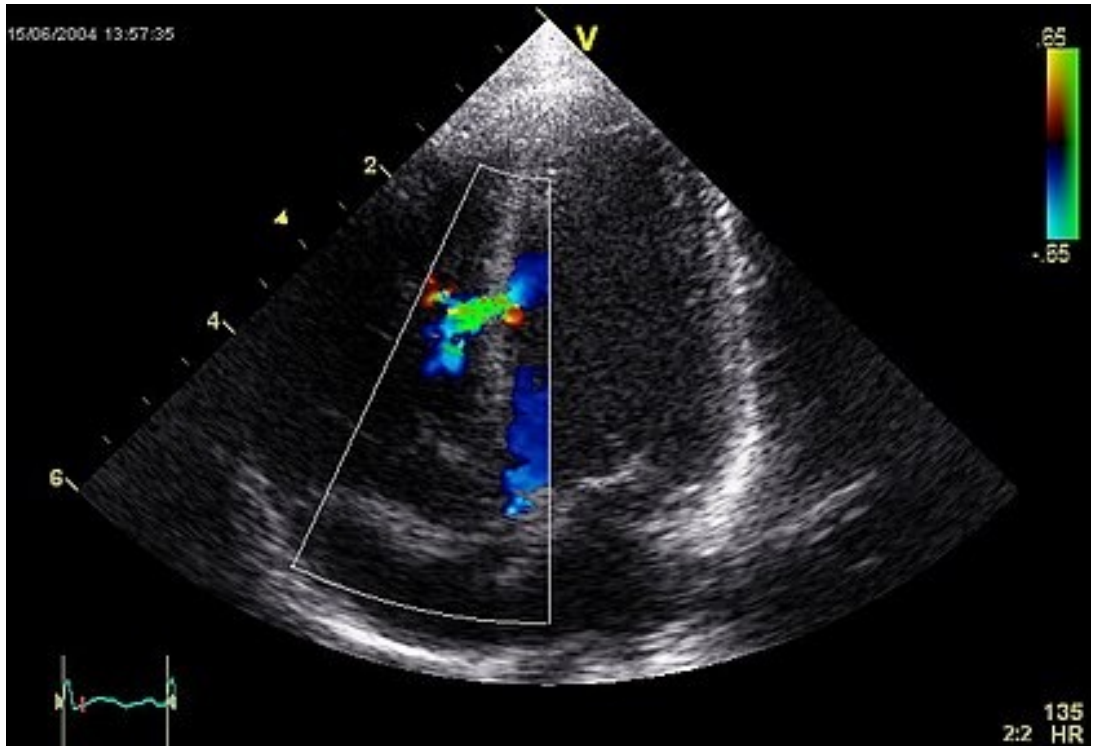
Cardiac ultrasound and Doppler (echocardiogram).

- Cardiac ultrasound gives a non-invasive dynamic picture of structure of the heart.
- Includes muscle wall, valves, pericardial sac.
- Doppler measures flow, and pressure across valves.
- Mainly transthoracic (probe on the chest wall).
- Can be transoesophageal, stress, (rarely) intracardiac.

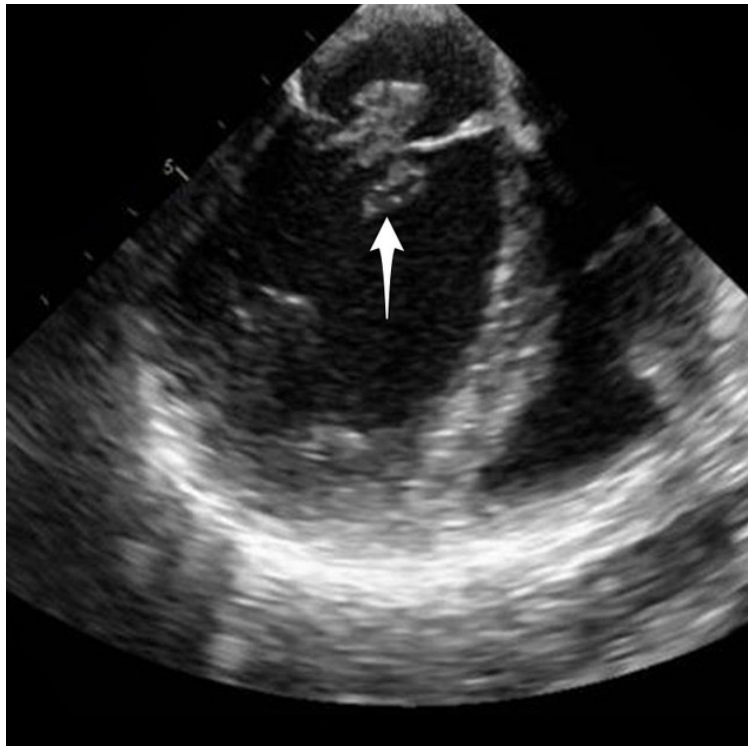


Patrick J. Lynch, C. Carl Jaffe

Echocardiography has transformed diagnosis of structural abnormalities of the heart.

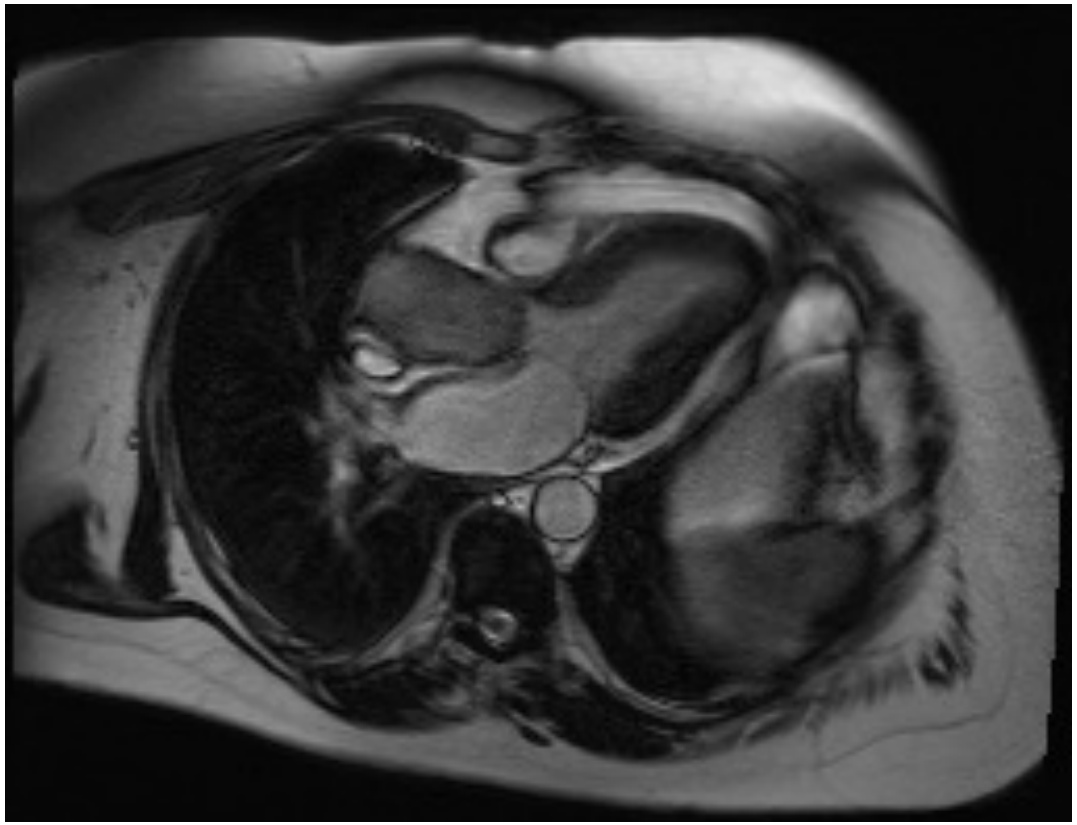


Atrial septal defect. Kjetil Lenes.



Mitral endocarditis. Frank Gaillard, Radiopaedia.

Cardiac MRI and CT used in complex cases.



Bicuspid valve, MRI. Sajoscha A. Sorrentino, Radiopaedia

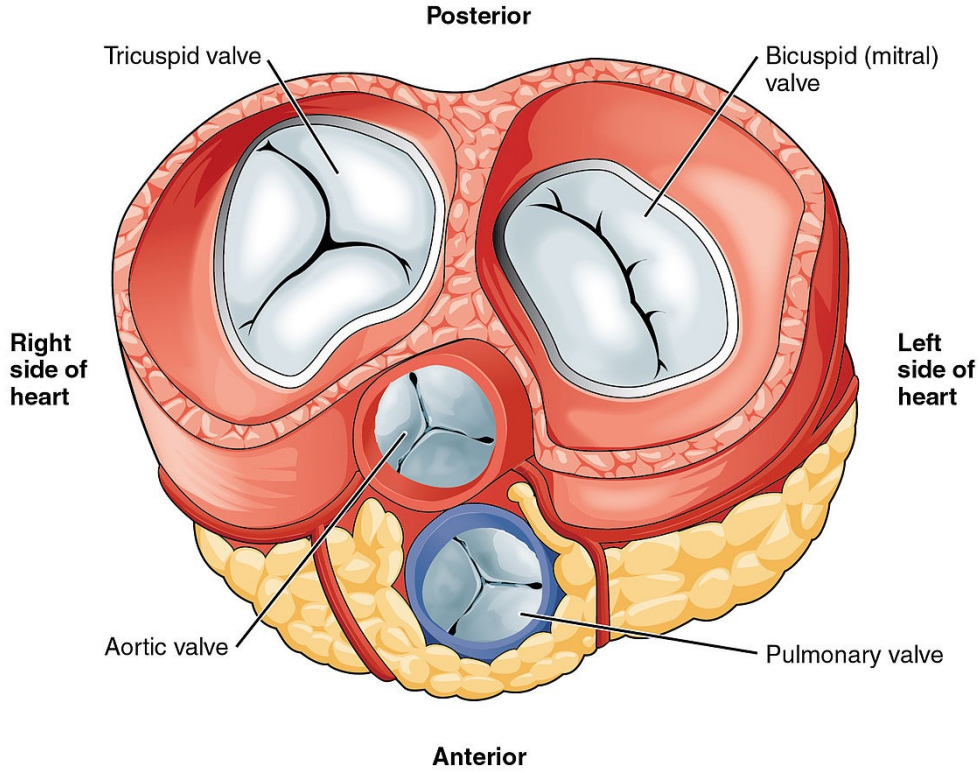
Aortic stenosis, CT, Frank Gaillard, Radiopaedia

Valves of the heart- stenosis and regurgitation.

- Left side:
- Mitral valve (atrium to ventricle)
- Aortic valve (ventricle to body)

- Right ride:
- Tricuspid valve (atrium to ventricle)
- Pulmonary valve (ventricle to lung)

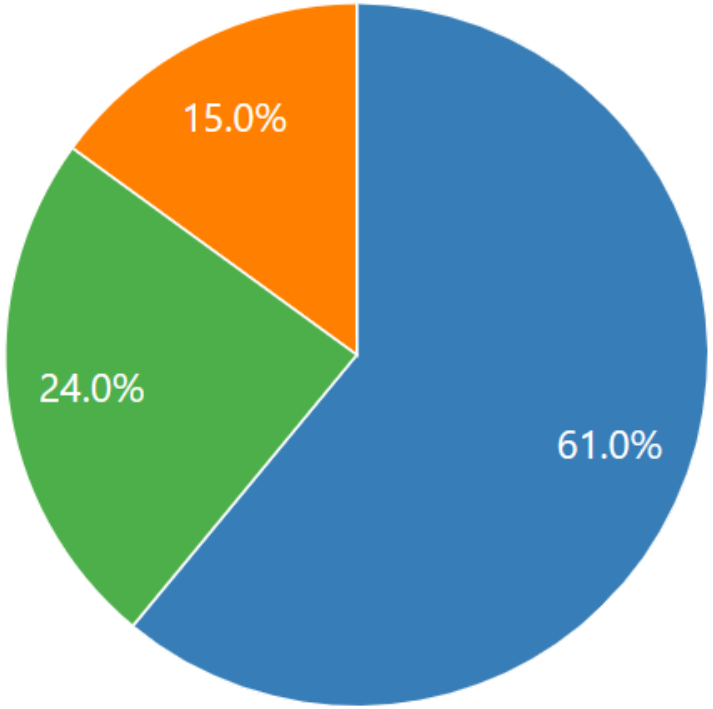
- Narrow (stenosis) or floppy / partially open (regurgitation).



Openstax College / Wiki

Valves are affected in valvular disease in adults.

- Clinically important disease mainly on the left side of the heart.
- Deaths from valvular heart disease, US (CDC) data.
- Aortic valve- about 61%
- Mitral valve- about 24%
- Other- about 15%

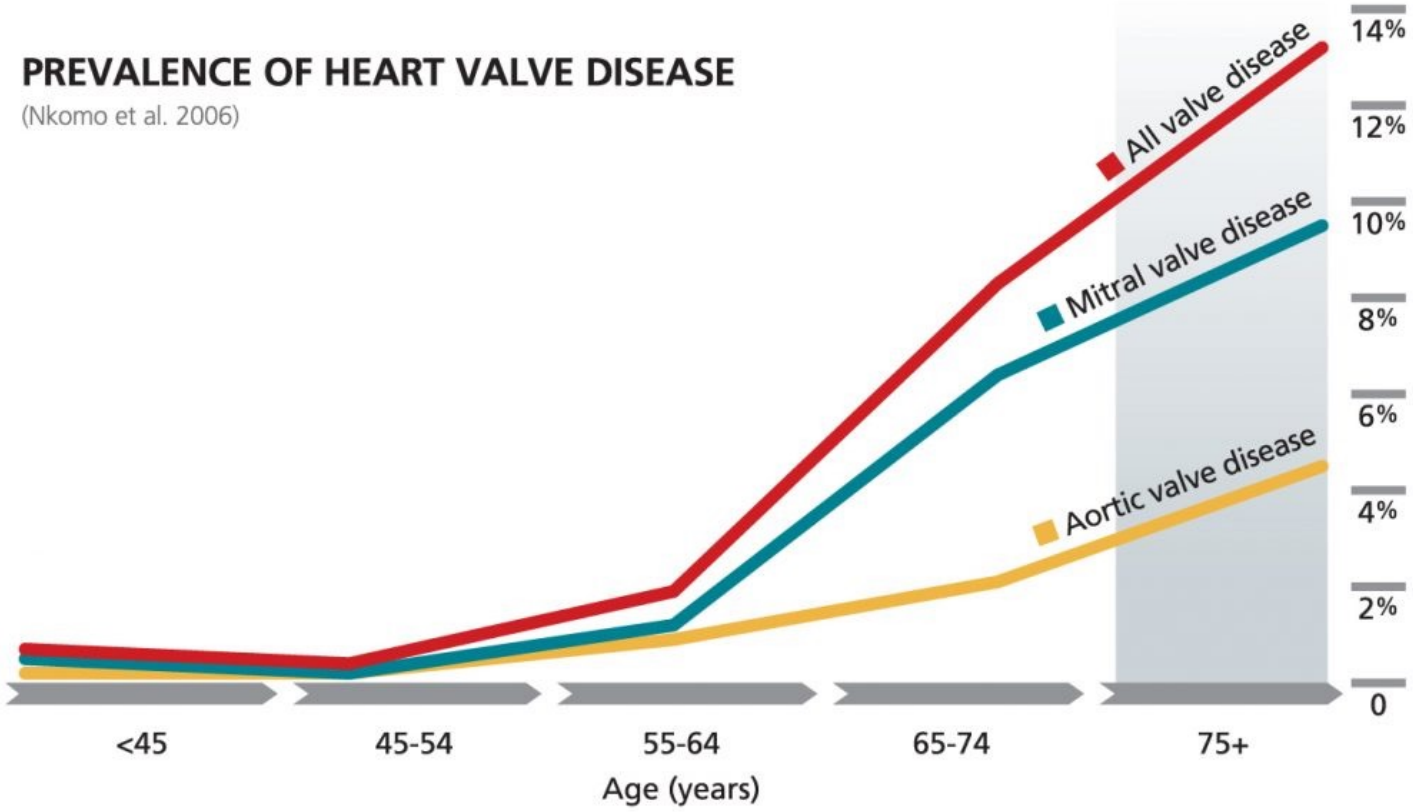


US CDC 2023

Minor stenosis or regurgitation prevalence increase with age. Degenerative-calcific in the great majority.

- Risk factors include:

- High lipids
- Hypertension
- Diabetes / obesity
- Chronic kidney disease
- Smoking.



Nkomo et al Lancet 2006, adapted by Silver Books

Symptoms of valvular heart disease.

- Many have no symptoms at all.

Symptoms include:

- Shortness of breath doing something used to
- Dizziness, fainting (esp. on exercise)
- Chest pain
- Fatigue
- Swelling of limbs or abdomen.

Peeples Gary, U.S. Fish and Wildlife Service

Bacterial and fungal endocarditis- infection on the valve.

- Around 1:30,000 get endocarditis.
- Risk factors:

Heart structural abnormalities.

-Artificial valve.

-Congenital heart abnormality.

-Hypertrophic cardiomyopathy.

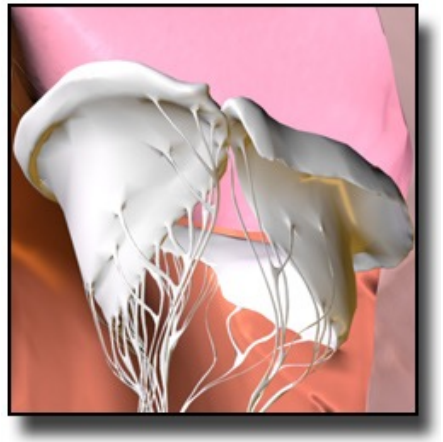
Risks for bacteria getting into blood.

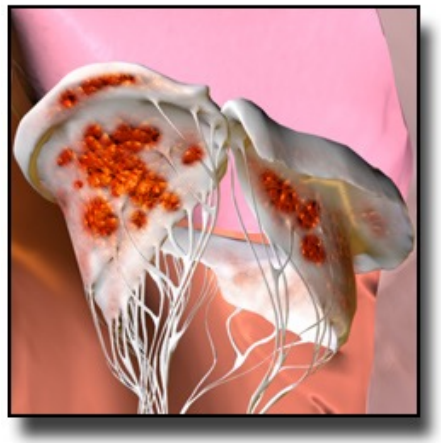
-Intravenous lines in hospital.

-Intravenous drug use.

-Dentistry / poor dentition.

-Surgery.





Bruce Blaus.

What organism, and treatment, depends on setting.

- Endocarditis can cause just fever, or emboli can flick off. Can cause strokes.
- When cause in hospital usually skin- Staphylococcus.
- In the community often dental- *Streptococcus viridans* group.
- From gut- *Streptococcus bovis.*
- Immunosuppressed- fungal (Candida).
- Treatment prolonged antibiotics, 20% surgery.

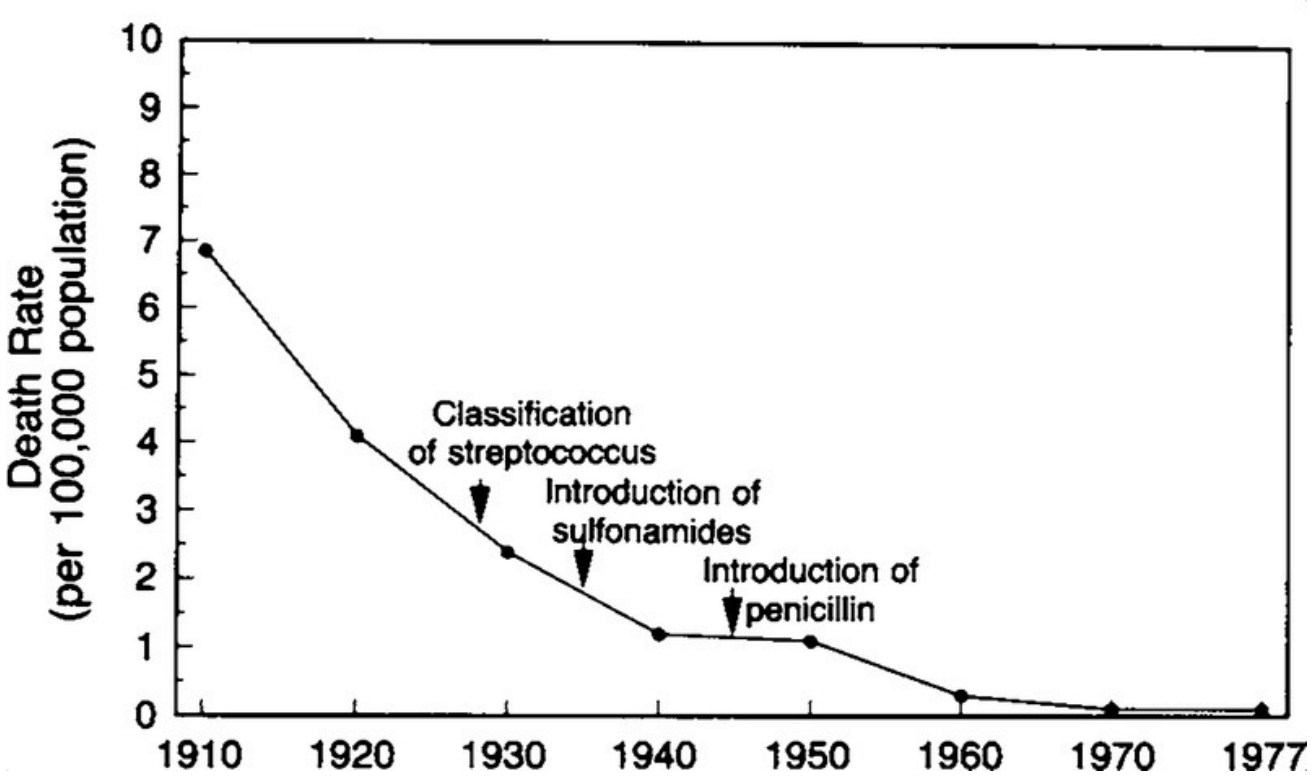
Splarka

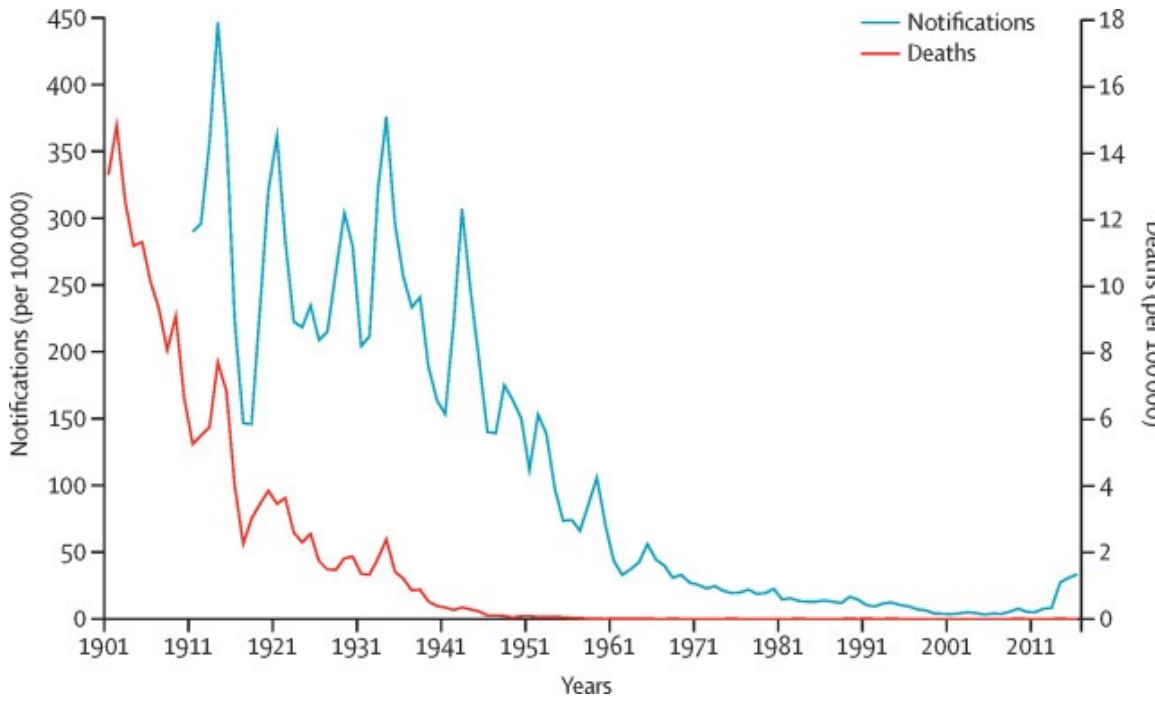
Acquired valve heart disease in childhood. Rheumatic heart disease.

- Rheumatic fever follows a Group A streptococcal throat or skin infection. Around 3% of untreated.
- Was very common in UK. Chorea 2nd/4th most common cause for admission at Great Ormand St. in 1880s.
- Immunological reaction.
- Effects throughout body (esp. joints).
- Early heart inflammation, up to 60% have long term heart problems.
- Damage to mainly mitral heart valve.
- Heart failure, atrial fibrillation.

St. Vitus's Dance / Sydenham's chorea

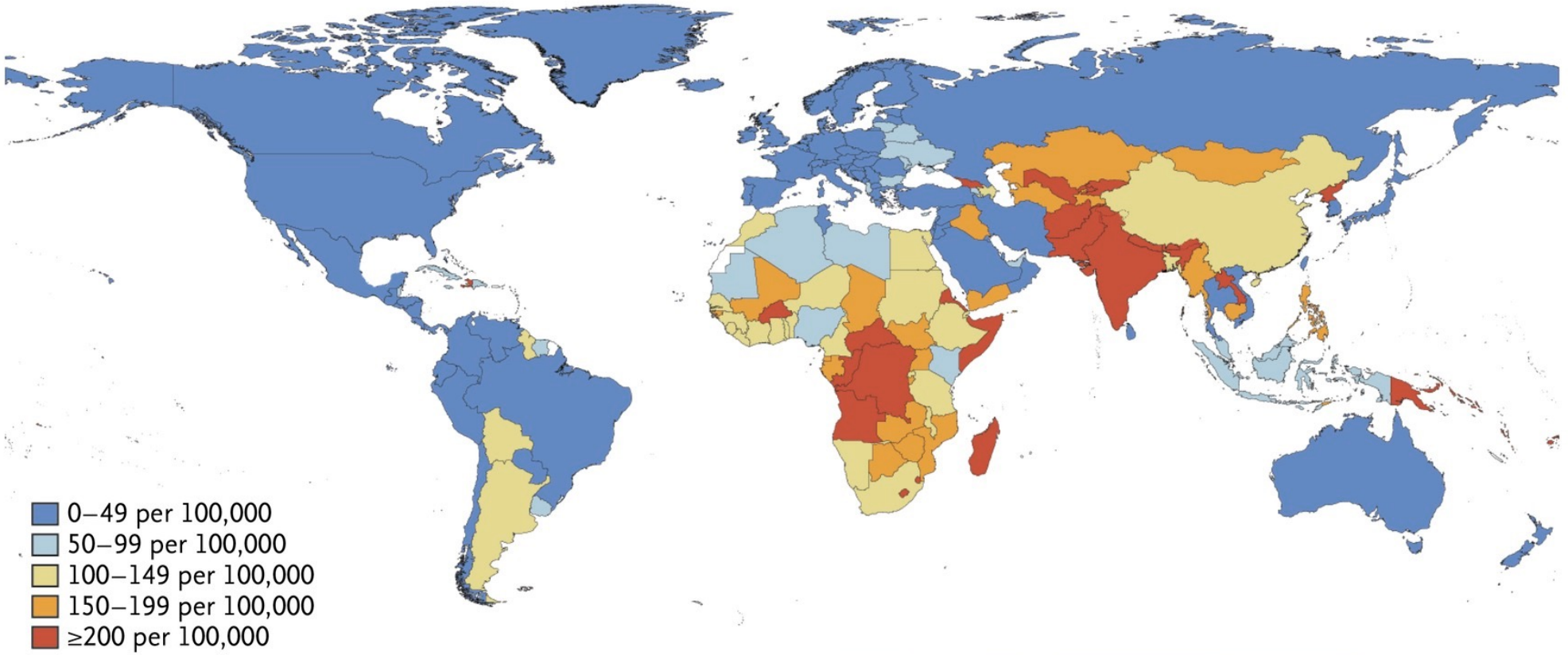
Deaths /100,000 from rheumatic heart disease over time. USA 1910-1980; Scarlet fever in UK 1901-2016.





Gordis L; Lamagni T et al Lancet ID 2018

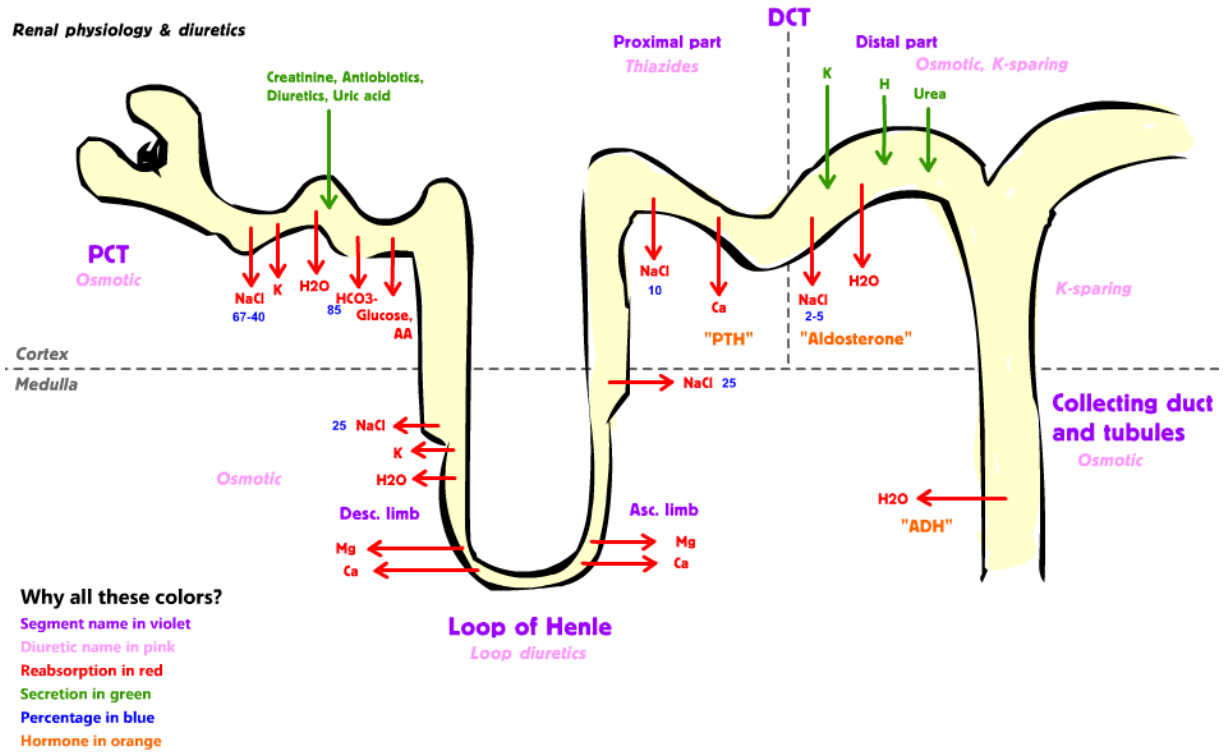
About 30 million affected by rheumatic heart disease globally, around 300,000 deaths p.a. (WHO). Strong link to poverty.



Age-Standardized Disability-Adjusted Life-Years Due to Rheumatic Heart Disease per 100,000 Population, 2015.
Watkins D et al NEJM 2017, GBDS.

Treatment of symptomatic valvular disease may be with drugs.

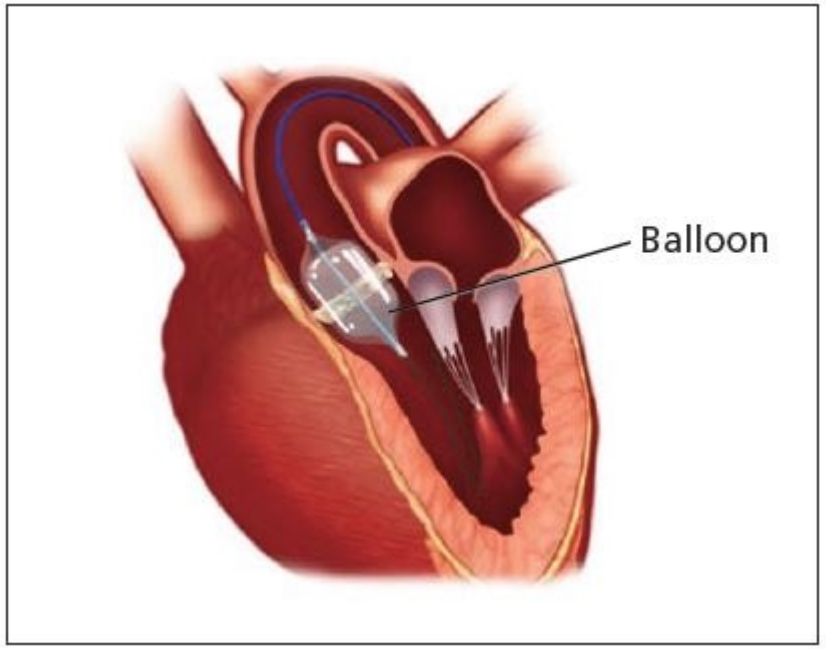
- Drug treatment does not cure valvular heart disease but may control the symptoms. Several drug classes including:
- Rhythm control with digoxin and other anti-arrhythmic drugs.
- Blood pressure control with nitrates and beta-blockers.
- Diuretics to reduce fluid build-up.
- Furosemide, thiazides.



Points of action of diuretics on the kidney. Haisook/Wiki

Severe acquired valve disease may need surgery.

- Wherever possible surgeons will use valve repair rather than valve replacement.
- More common in mitral than aortic valve disease.
- This has the advantage it is your native valve- less risk of clots or infection.
- The most minimally invasive is a balloon aortic valvoplasty.
- Balloon on a catheter inflated in a stenosed aortic valve to stretch it.
- Often only has an effect for a year.



Royal Brompton and Harefield Hospitals. NHS

Most valve repair and replacement will still require open heart surgery.

- Standard method requires the sternum (breastbone) to be split.
- The circulation on a bypass machine (heart-lung machine).
- The heart is stopped temporarily.
- Surgeon repairs or replaces the heart valve.
- The heart is restarted.

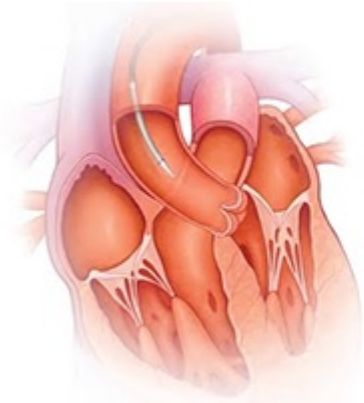
Valve replacement, Kiev 2019, Lena Gulenko.

Valve replacement and artificial heart valves.

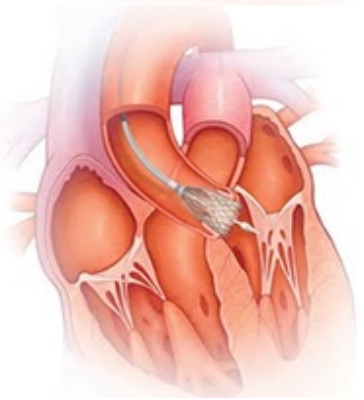
- These have evolved from large ball-and-cage valves.
- Various tilting disk valves.
- Tissue valves- fibrous, or from humans (cadaveric), or occasionally from animals (eg pigs).
- Which is best depends on age among other factors.

Z22

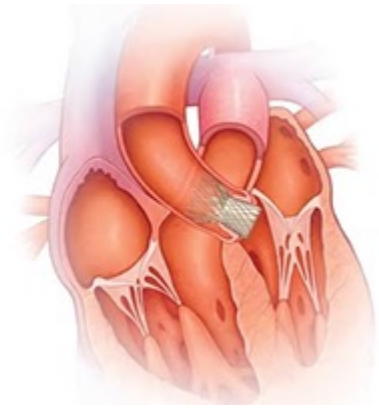
Less invasive surgical repair or replacement.



- Some centres now use less invasive surgical methods with several cuts to the chest and leg.



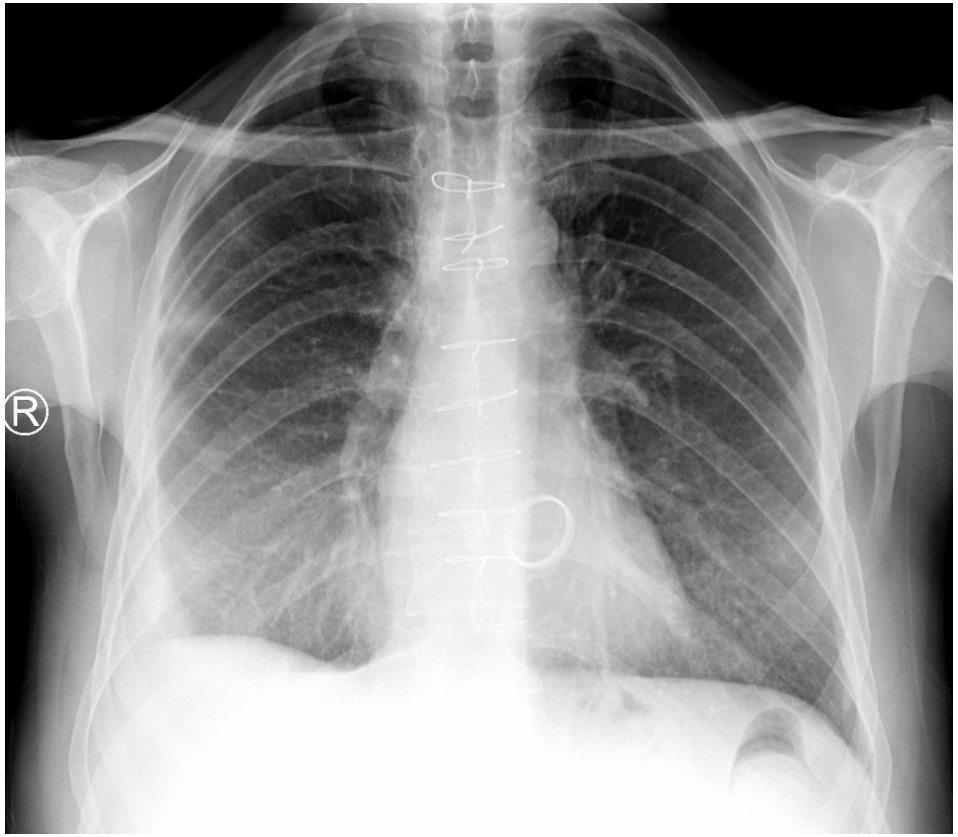
- For patients who are at high anaesthetic risk, a transcatheter aortic valve implantation (TAVI) procedure for aortic stenosis.
- A catheter is inserted into the groin and advanced to the heart. A valve fixed over the existing valve.
- Can be done under local anaesthetic.



Leeds Teaching Hospital NHS Trust

Valve surgery has been transformative.

- Many people who would have died, or lived very constrained lives have quality and quantity of life significantly improved.
- This is major surgery with risks as well as benefits.
- Includes risk of stroke during or soon after operations.
- Cardiac bypass puts considerable strain on the body.

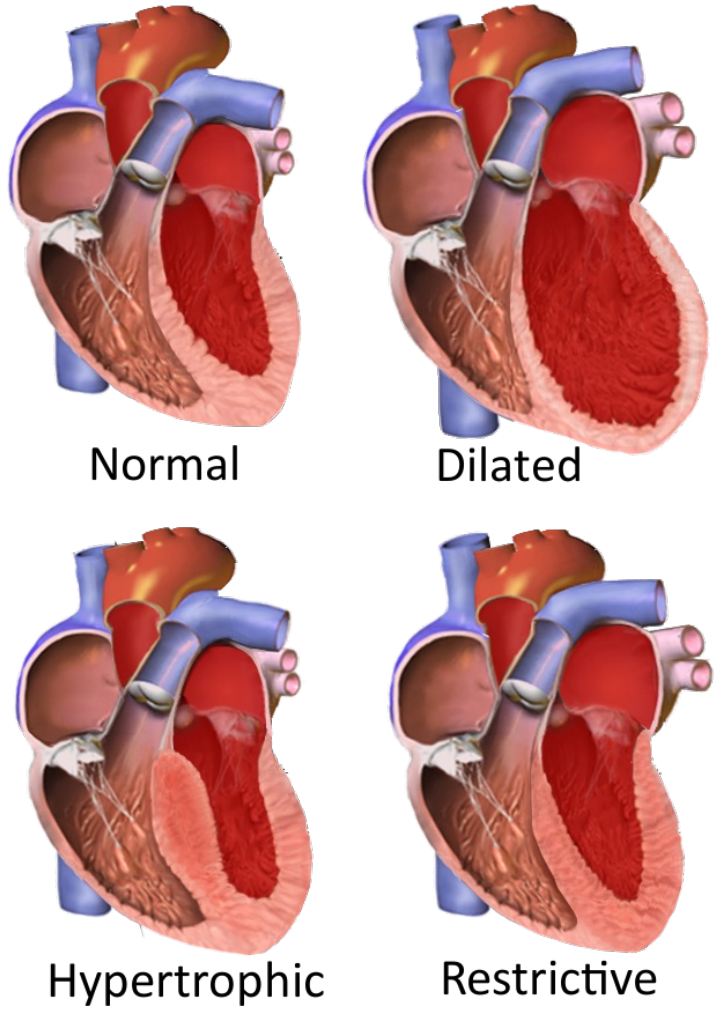


Mistral valve replacement.
Dr. Henry Knipe, Radiopaedia

Cardiomyopathy- damage to the heart muscle.

Three main types:

- Dilated cardiomyopathy. Most common.
- Hypertrophic cardiomyopathy- usually inherited.
- Restrictive cardiomyopathy.



Npatchett, BruceBlaus

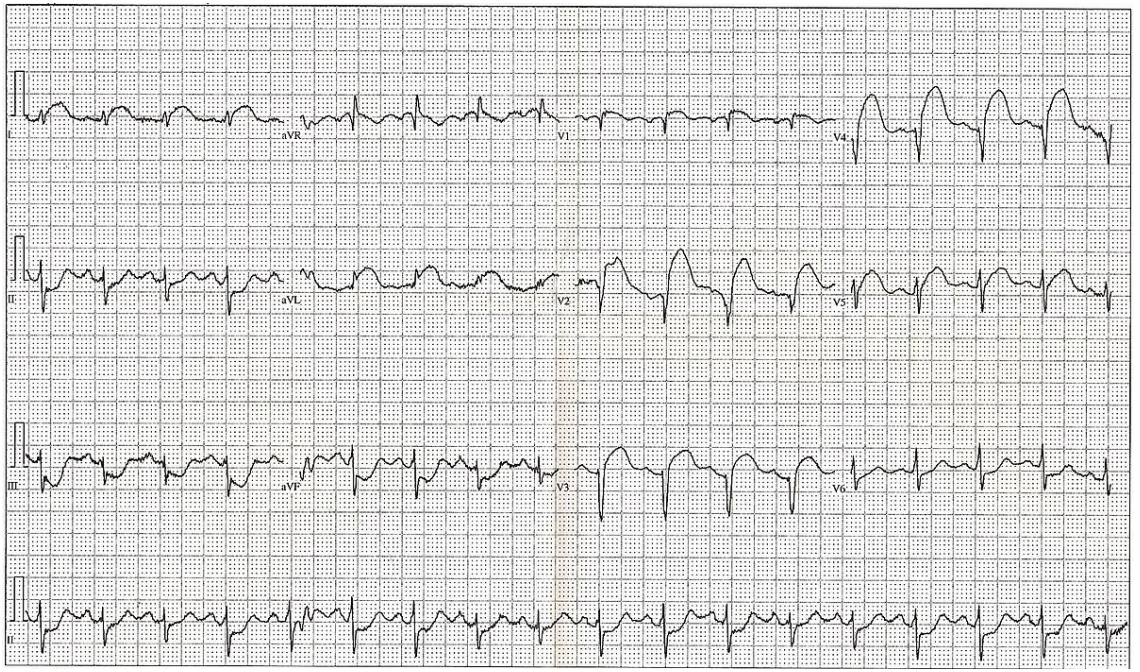
Common symptoms of cardiomyopathy.

- Shortness of breath, especially on exercise.
- Fatigue.
- Dizziness, light-headedness on exercise.
- Swelling (oedema) of feet, legs.
- Arrhythmia.
- Chest pain.
- A new heart murmur.

Metropolitan Museum of Art.

Ischaemic cardiomyopathy- damage to heart muscle.

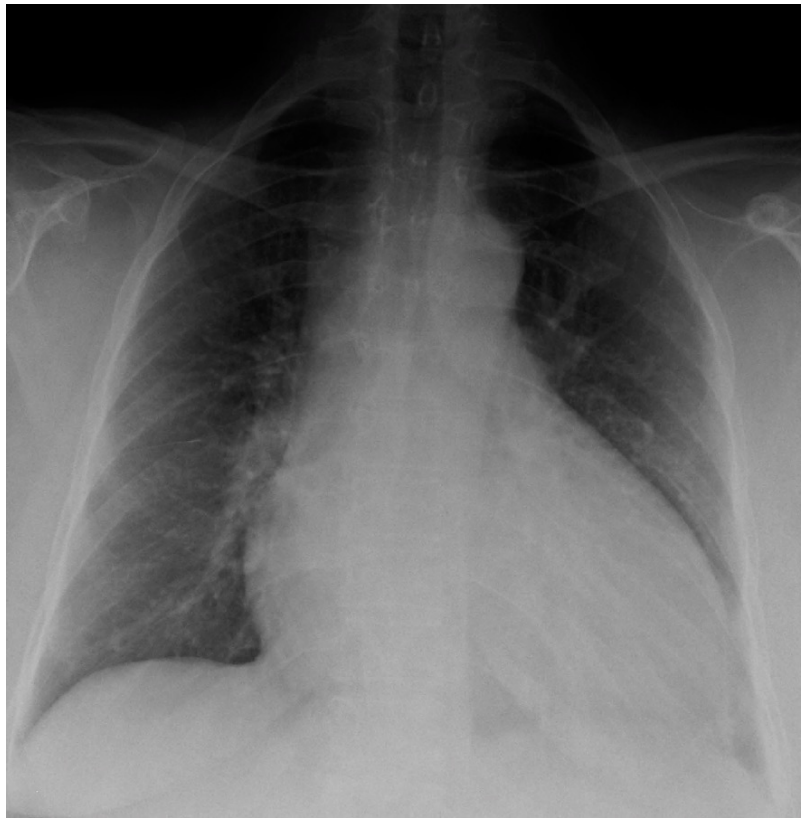
- The commonest cause of dilated cardiomyopathy in high-income countries is ischemic.
- Caused by heart attacks and other ischaemic damage to the heart muscle.
- Common cause of heart failure.
- Prevention and early treatment same as for coronary heart disease.
- Hypertension independent risk factor for cardiomyopathy.



Wiki

Drugs, toxins and dilated cardiomyopathy.

- Several dugs and toxins can damage the heart muscle.
- Alcohol in large quantities over time- alcoholic cardiomyopathy.
- Cocaine and amphetamines.
- Some chemotherapy drugs may.
- Some toxins and chemicals used in industrial processes- lead, mercury, cobalt.



James Heilman

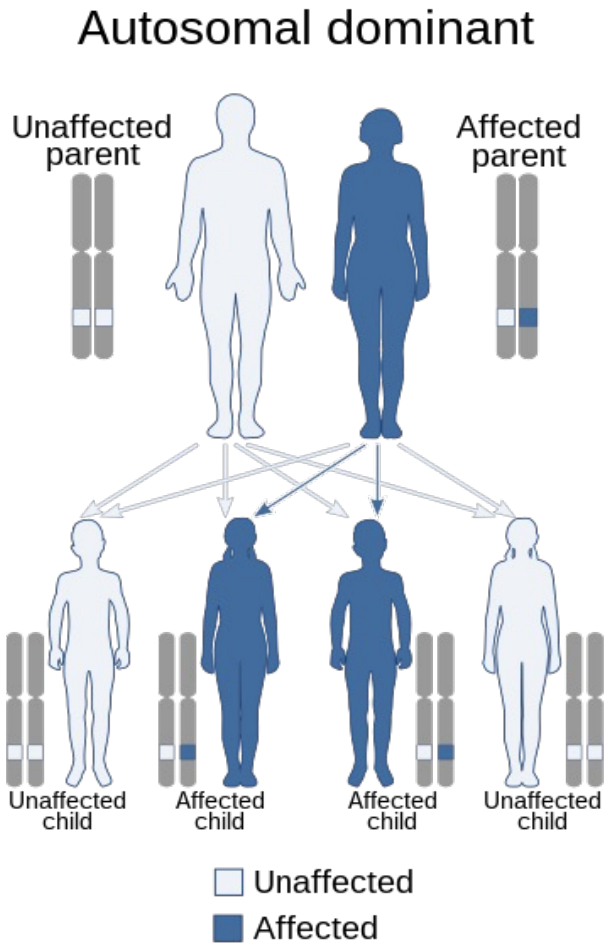
Dilated cardiomyopathy during or after pregnancy.

- Peripartum cardiomyopathy rare- but important. 1:1300-4000 live births.
- Occurs late in pregnancy or in the first few weeks after delivery.
- Needs to be identified and treated early. Shortness of breath, swelling (oedema).
- Over 50% recover completely but this may be slow.
- Cause unknown.

Jacob Sippel

Hypertrophic cardiomyopathy (HCM).

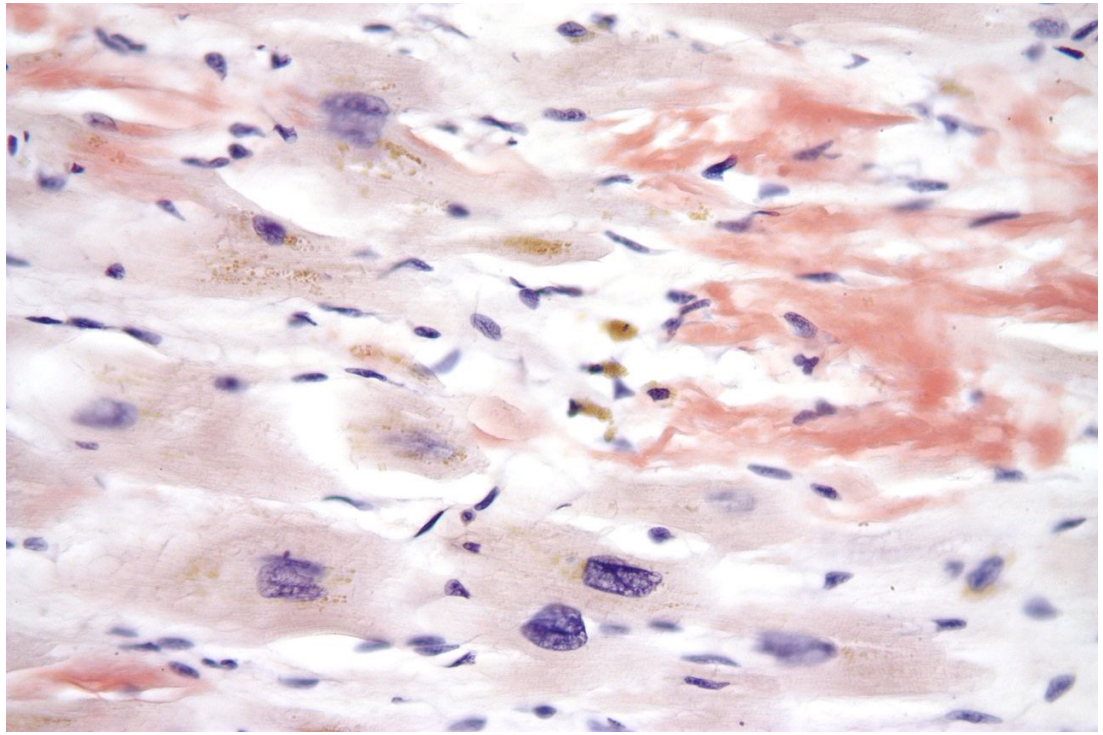
- Can occur any age but most commonly begins late teens- early adults.
- Usually inherited. Maybe 1:500, most undiagnosed.
- All first-degree blood relatives should be screened.
- Genes associated with the sarcomere. Autosomal dominant. Many are not known.
- Can be associated with sudden deaths, especially athletes- but very rare. 1 death per 220,000 athletes.
- About 1% a year mortality risk- but many have no symptoms and normal lifespan.



SUM1, Domaina, Angelito7

Restrictive cardiomyopathy.

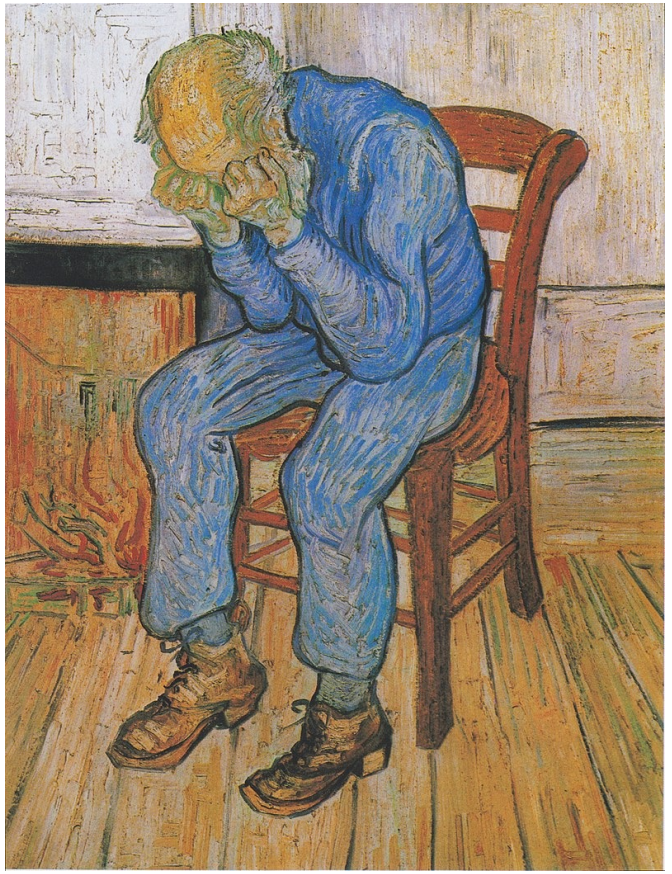
- Heart muscle becomes more rigid, less able to contract.
- Can be caused by infiltration or scarring.
- Haemochromatosis- too much iron in the body.
- Sarcoidosis.
- Amyloid.
- Connective tissue diseases.
- Some drugs and radiation.



Amyloid infiltration of heart muscle.
Congo red. Nephron.

Stress-induced cardiomyopathy / broken heart syndrome.

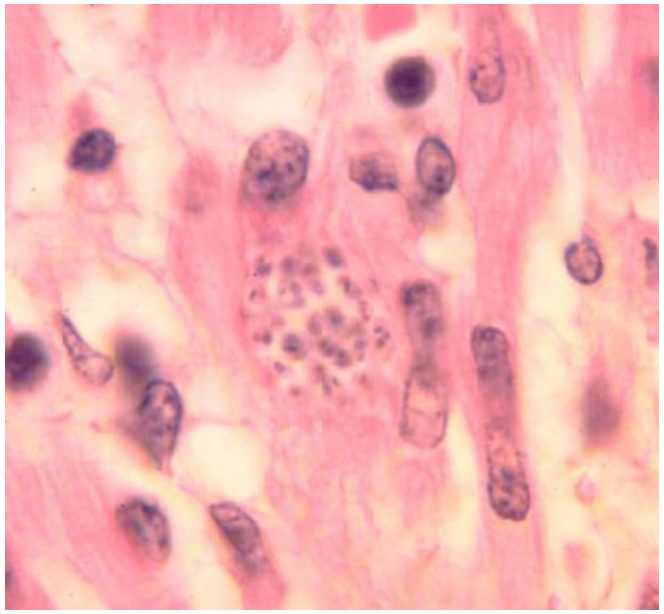
- Sudden myocardial dysfunction after a stressful event such as a bereavement or divorce.
- Chest pain, shortness of breath, abnormal heart movements, abnormal blood tests and ECG.
- Can lead to severe heart dysfunction- but temporary (days to weeks).

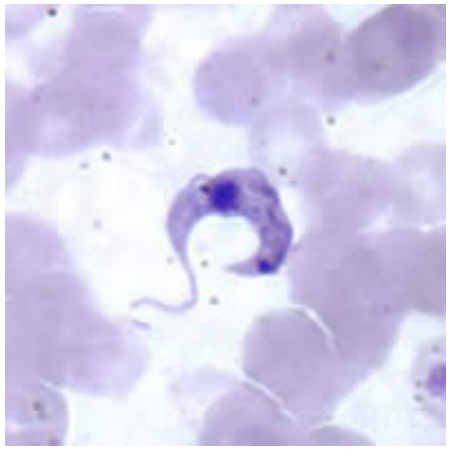


Vincent Van Gogh. Op de drempel van de eeuwigheid. 1890

Chagas cardiomyopathy.

- Chagas disease is endemic in large areas of rural Latin America.
- A trypanosome parasite passed on by reduviid bugs.
- Still the most common cause of non-ischaemic cardiomyopathy in Latin America.
- 5.7 million people in 21 countries have Chagas (WHO); 20-30% will get cardiomyopathy in 20 years.
- Substantial reduction in new cases.

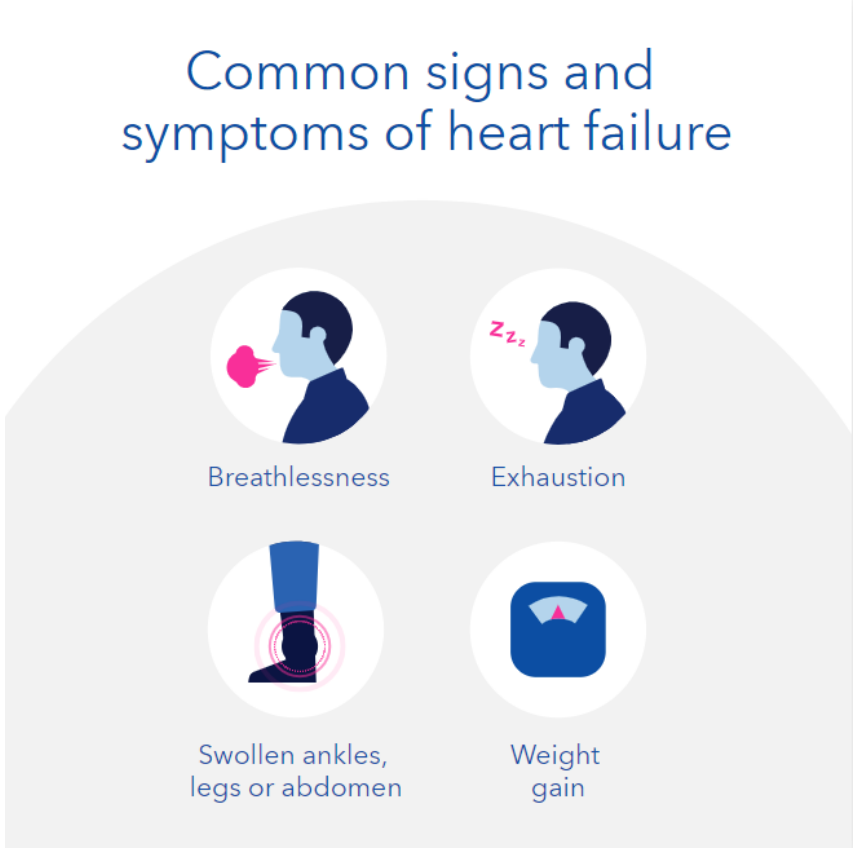




Wiki

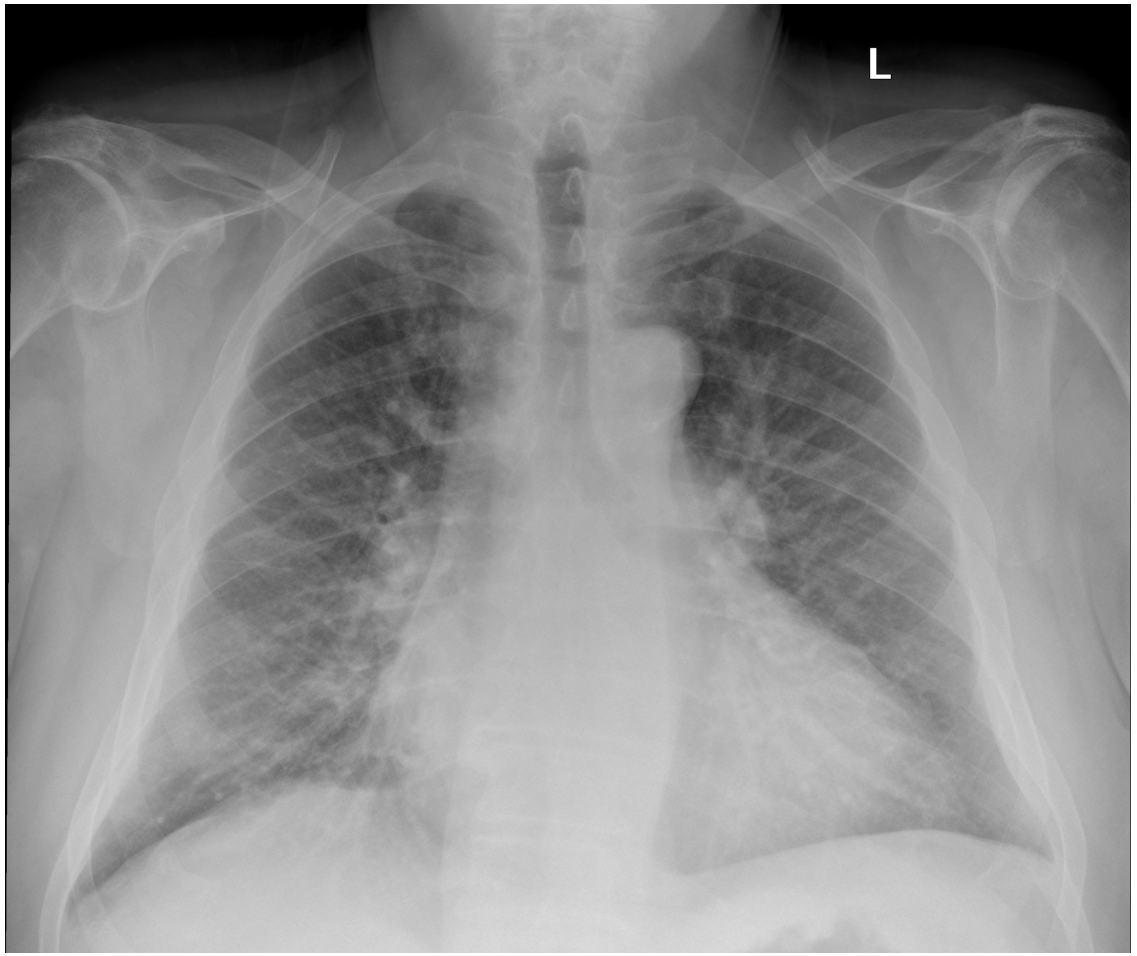
The end result of cardiomyopathy is likely to be heart failure. Not all heart failure is due to cardiomyopathy.

- Can be from mild to severe. Staging of severity by NYHA class:
- class 1 – you don't have any symptoms during normal physical activity
- class 2 – comfortable at rest, normal physical activity triggers symptoms
- class 3 – comfortable at rest, minor physical activity triggers symptoms
- class 4 – unable to carry out any physical activity without discomfort and may have symptoms even when resting.



NHS Manchester University Foundation Trust

Left heart failure fluid accumulates in lungs- shortness of breath.
Right heart failure- fluid accumulates in peripheries- swelling.

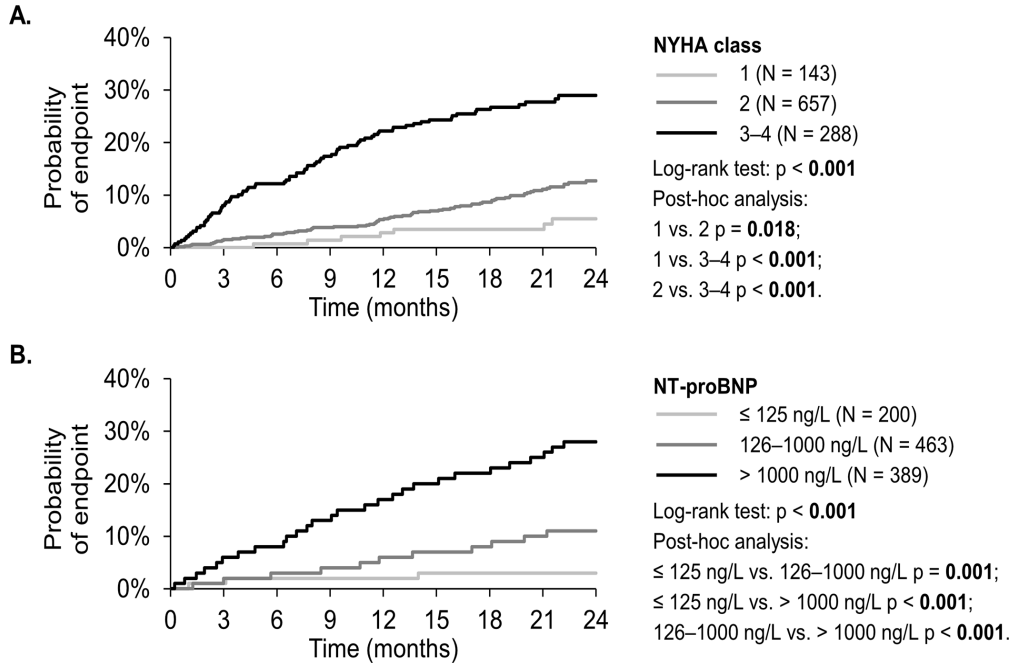


Congestive cardiac failure. Frank Gaillard, Radiopaedia

Pitting Oedema. James Heilman.

Treatment of heart failure depends on severity.

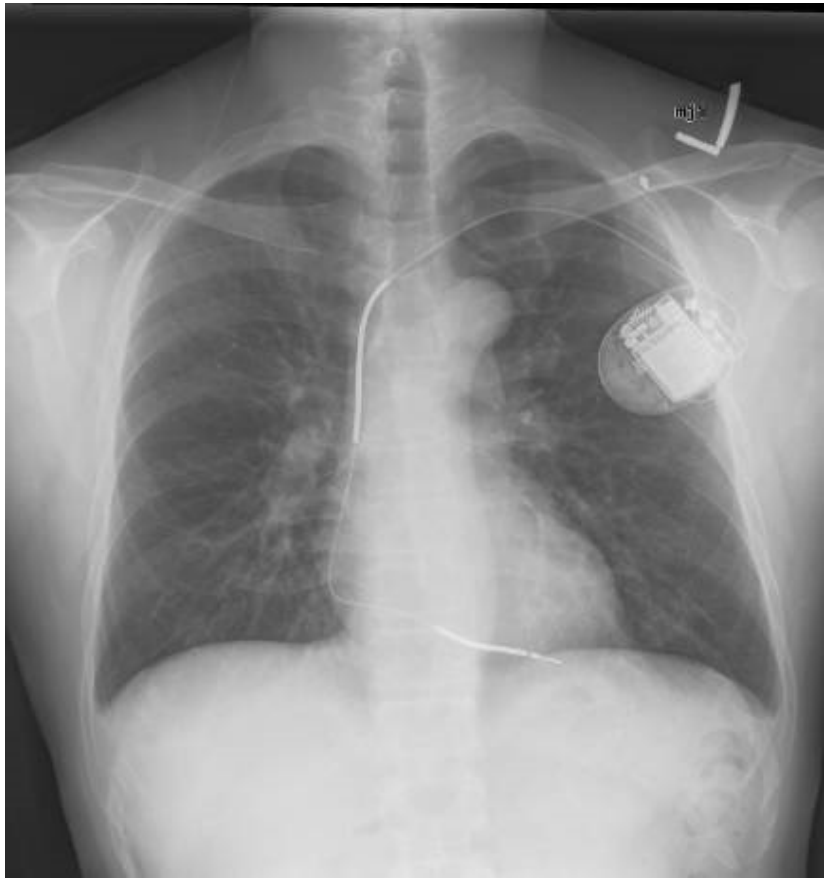
- Usually confirm diagnosis with N-terminal pro-B-type natriuretic peptide (NT-proBNP) blood test, echocardiogram.
- Modify risk factors where possible to slow progression- smoking, blood pressure, cholesterol. Depends on cause.
- ACE inhibitors and Beta-blockers first line treatment. ARBs (angiotensin II inhibitors) if do not tolerate ACE inhibitors.
- Mineralocorticoid receptor antagonists (MRA) such as spironolactone if still have symptoms.



Endpoint death, transplant/LVAD
Spinar J et al PLOS One 2019

In more severe cases a range of drugs and procedures considered to optimise remaining cardiac function.

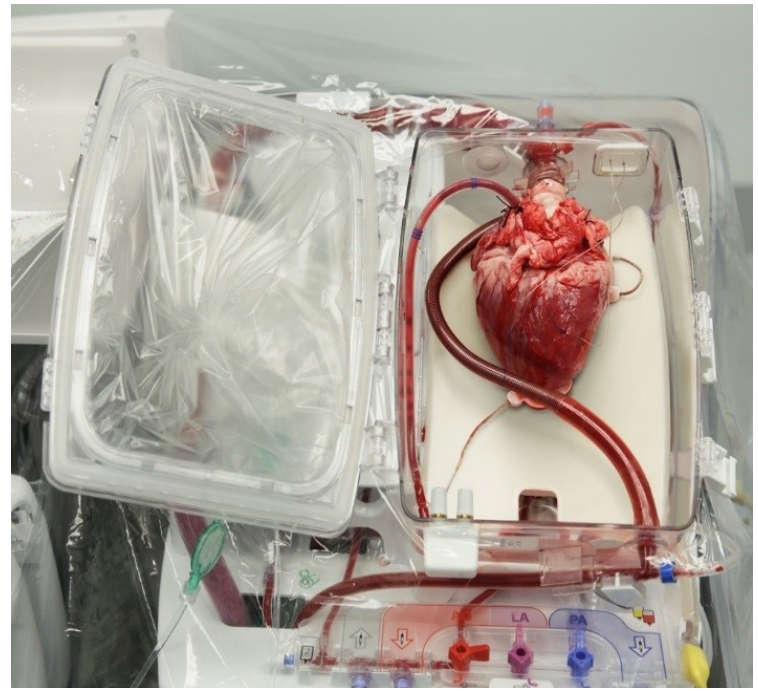
- In severe (class 2 and 3) cases specialist clinics may use a range of drugs in combination.
- Getting the balance of drugs right for an individual often takes time.
- Pacemakers, cardiac resynchronisation therapy (CRT) devices implantable cardioverter defibrillators (ICDs) to optimise rhythm.
- Coronary artery stents, and coronary artery bypass surgery if IHD.



ICD device. Gregory Marcus.

Heart transplant and left ventricular assist devices.

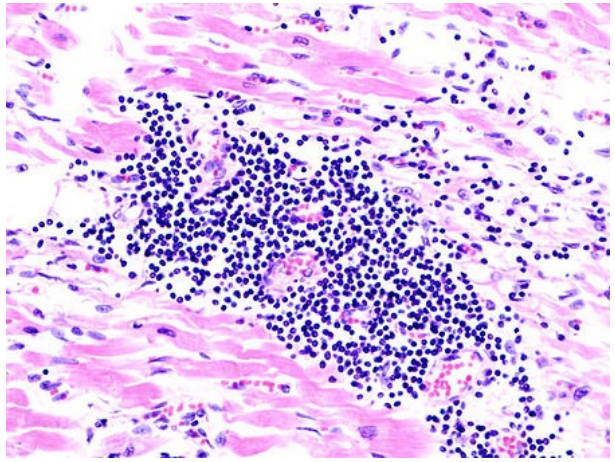
- Very small numbers. Major surgery.
- Left ventricular assist devices (LVAD) an artificial pump into the heart. About 80 a year in the UK.
- Heart transplants around 100-150 a year. Needs a good match.
- Many risks, including graft rejection, graft failure, reaction to drugs. But:
- 70 to 75 in every 100 people with a transplant will live at least 5 years (NHS).
- 50 / 100 people will live at least 10 years.



Korozia45

Viral and bacterial myocarditis.

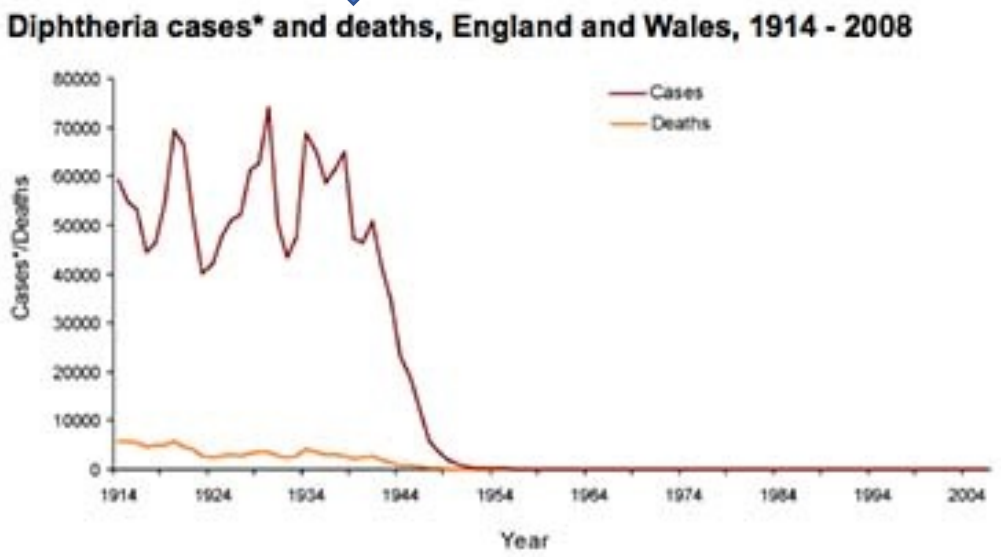
- In 2017, in the UK it was estimated that there were about 2,000 hospital admissions for myocarditis.
- Most recover. Important cause of heart disease in young adults, children; occasional sudden deaths.
- Viral most common. Include Coxsackie B, Parvovirus B19, HIV, Hepatitis C, COVID-19.
- Bacteria include Haemophilus, legionella, mycoplasma, brucellosis, Q-fever, typhus.
- Initial inflammation due to infection, then in some a second immune inflammation.



Heart inflammation. Wiki.

Diphtheria and the heart- toxin meditated myocarditis.

- Used to be a major cause of death, especially in children.
- The toxins of diphtheria cause myocarditis.
- Can cause heart block.
- Diphtheria now rare; better living conditions, antibiotics, vaccination (UK from 1942).
- In 2015, 4,500 cases were officially reported worldwide; over 1m before 1980s. Fatal in 5-10%.



Non-infectious myocarditis.

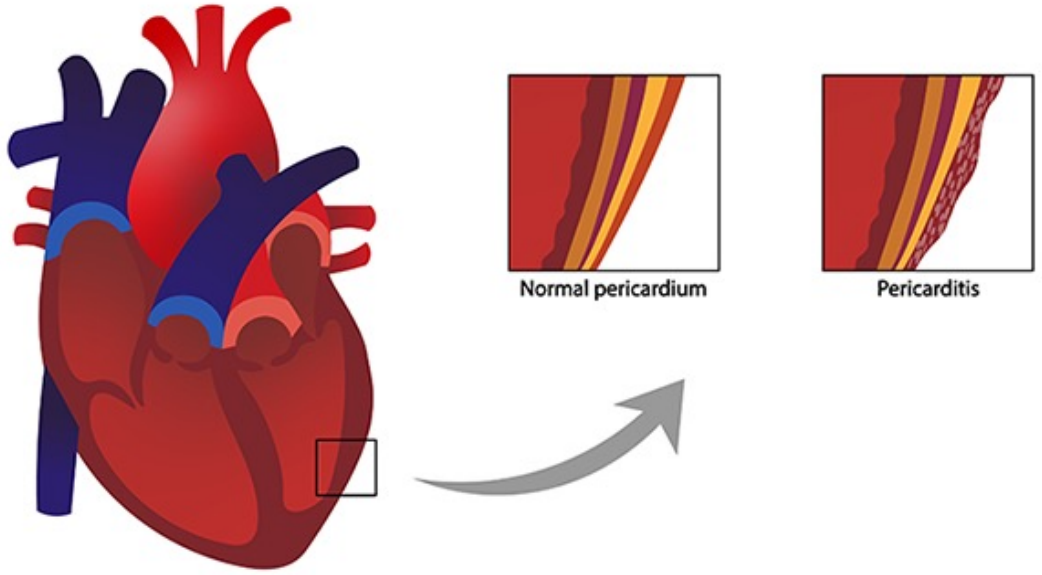
- Inflammatory conditions.
- Autoimmune conditions including sarcoid, lupus.
- Some drugs and vaccines.
- Some environmental toxins.
- All rare.

- Most myocarditis is a one-off event, and people recover after rest.

Lupus erythematosus. Atlas der Hautkrankheiten 1856.

Pericarditis- inflammation of the covering of the heart.

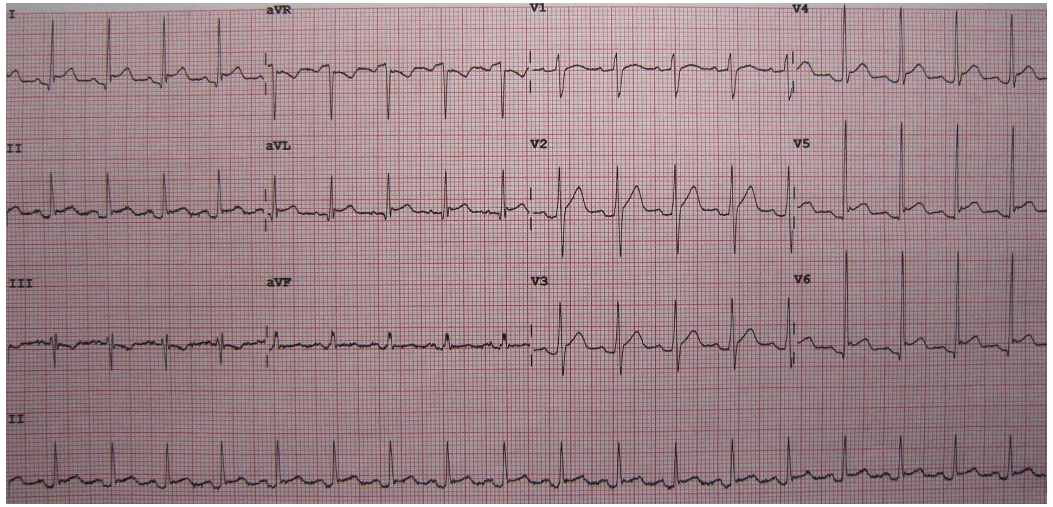
- Chest pain, often sharp, stabbing.
- Often worse lying down or taking a deep breath.
- Made better by leaning forward
- ECG changes.
- Usually acute pericarditis gets better on its own in a few days to weeks, occasionally months.
- Rarely dangerous on its own but some exceptions.
- Can cause rhythm disturbances.
- Often occurs with myocarditis.



BHF

Causes of pericarditis include:

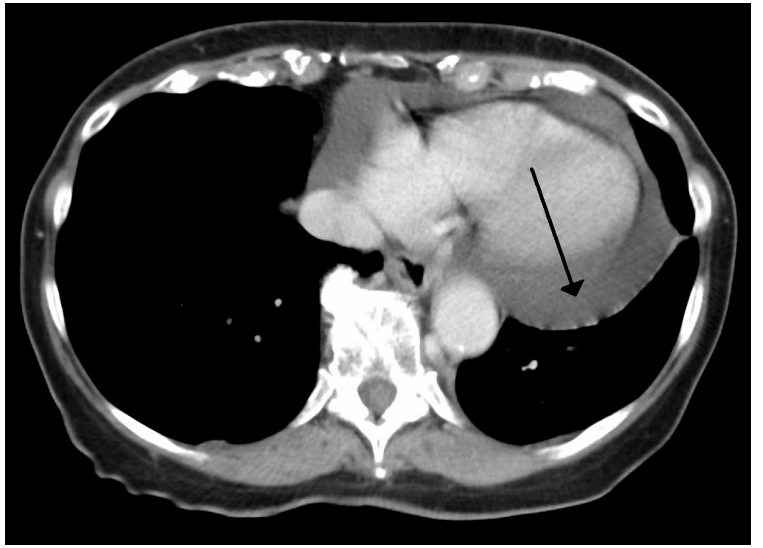
- Most commonly a viral infection- includes ‘flu, COVID-19.
- Other infections including bacterial, tuberculosis, fungal.
- Inflammatory diseases like rheumatoid arthritis.
- Heart attack or heart surgery.
- Drugs and vaccines- includes COVID-19 but at a much lower rate than in the infection.
- Blunt trauma to the chest.



ECG in pericarditis.
James Heilman MD

Pericardial effusion.

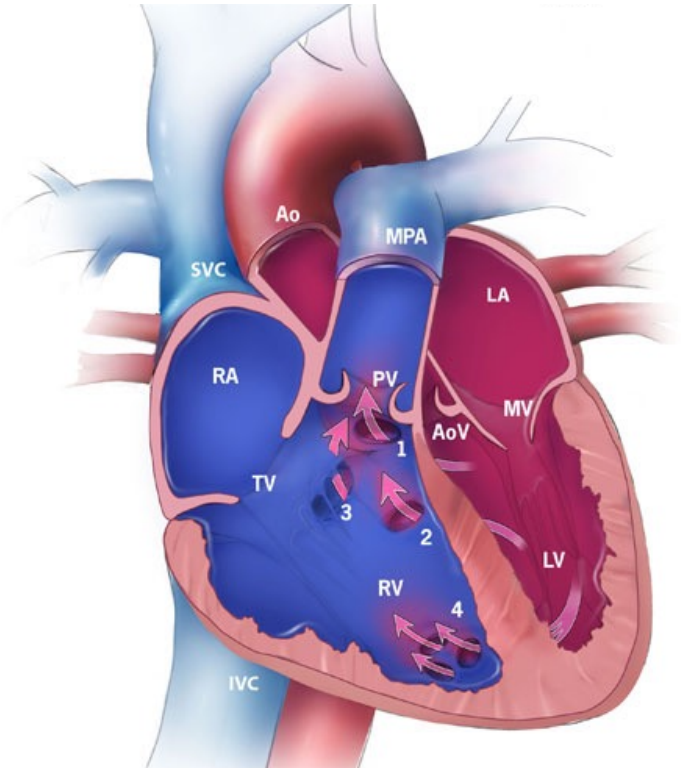
- Many infections, inflammatory conditions and other causes can cause a small pericardial effusion- fluid in the pericardial sac.
- Haemorrhage into the pericardial sac due to heart wall rupture after heart attack, surgery or trauma an emergency.
- The classical cause of a slower onset massive pericardial effusion is tuberculosis (TB). Other inflammatory causes and cancers can also cause it.
- Can squeeze the heart (tamponade).



J Heilman Wiki.

Congenital valve and heart structure conditions.

- Professor Martin Elliott delivered Gresham lectures on this (online).
- Some are common and relatively minor or self-correcting, some rare but serious.
- Just under 1% births in high-income settings (CDC data). Of these around 1:4 critical.
- Ventricular septal defect most common (‘hole in the heart’) 20%, then atrial septal defect.

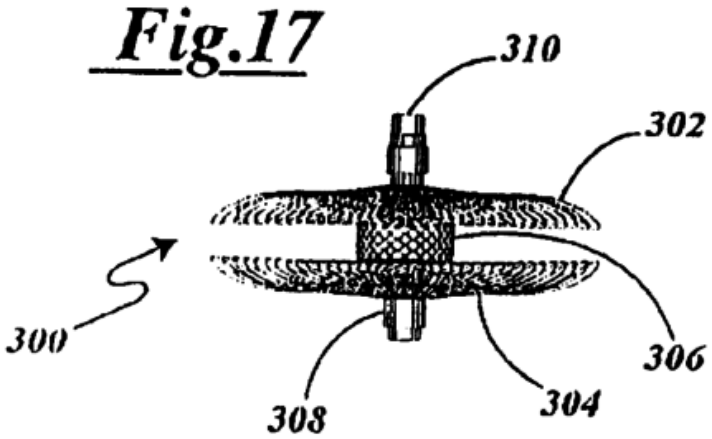


Ventricular septal defect.

CDC National Center on Birth Defects and Developmental Disabilities

Smaller congenital abnormalities often improve over time.

- Holes in the heart often self correct early in life or are minor and require no intervention.
- Patent ductus arteriosus (PDA) may close after drug treatment.
- Some larger holes in the heart can be closed via a catheter inserting a closure device.
- Valve abnormalities may need balloon valvuloplasty or implanted valve.
- A minority with more major cardiac abnormalities need surgery, sometimes in stages.



Amplatzer Septal Occluder (from the patent).

Three broad groups of things can lead to the heart failing.

- Myocardia ischaemia- lack of oxygen usually due to narrow or blocked coronary arteries (coronary heart disease).

- The heart rhythm is abnormal- either too fast or too slow (arrhythmia).

- The structures of the heart are damaged including muscle and valves.

Leonardo da Vinci. Circa 1511-13.