Heart Failure
Cardiomyopathy
Sudden Cardiac Death

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Heart failure

- **Broad descriptive term which includes:**
  - Left ventricular systolic impairment
  - Left ventricular diastolic impairment
  - Biventricular systolic impairment
  - Right ventricular systolic impairment

- **Prevalence**
  - approx 1% total population
  - 4-5% of population > 70 years
  - > 40,000 patients
  - Prevalence probably increasing as mortality from heart disease reduces
  - Most common discharge diagnosis in > 65s in USA
Heart Failure - Outcome

- Annual mortality is 15-20%
- From time of diagnosis 50% 5 year survival
- Morbidity high
  - Of those discharged from hospital with heart failure 30% will be readmitted in 3 months
Causes of Heart Failure

- Ischaemic heart disease
- Valvular heart disease
- Hypertension
- Inherited cardiomyopathy
- Congenital heart disease
- Infection
  - Myocarditis, endocarditis, rheumatic fever
- Pericardial disease
- Infiltrative (sarcoid, TB, haemochromatosis, metastatic cancers)
- Other:
  - Tachycardia
  - Peri-partum
  - COPD / OSA
Symptoms of heart failure

- Symptoms
  - Fatigue
  - Shortness of breath on exertion
  - Orthopnoea
  - Paroxysmal nocturnal dyspnoea
  - Palpitations
  - Dizziness, syncope
  - Angina
NYHA Classification of Symptoms

- **Class I** – no limitation of physical activity
- **Class II** – slight limitation of physical activity, comfortable at rest
- **Class III** – marked limitation of physical activity, comfortable at rest
- **Class IV** – symptoms at rest
Signs of left heart failure

- Gallop rhythm
- Lung crepitations
- Displaced apex beat
- Pleural effusions
- Signs of underlying cause
  - Valvular heart disease, hypertension etc
Signs of right heart failure

- Elevated JVP
- Ankle oedema (JVP should be up 4cms)
  - Cave venous insufficiency, Ca antagonist Rx
- Ascites
Diagnosis of Heart Failure

- Clinical history
- ECG
  - Often non-specific changes or fairly normal
  - Previous MI, acute ischaemic changes
  - Hypertensive changes or cardiomyopathy
  - Tachyarrhythmia
Echo in Heart failure

- LV size
- Systolic function
- Regional vs global wall motion abnormality
- Valvular heart disease
- LV diastolic function
  - Mitral valve inflow
  - Pulmonary vein flow
  - Tissue Doppler velocity
- Right heart size and function
Other investigations

- Chest X-ray
- Blood tests
  - BNP (pro-BNP, NTproBNP)
  - Anaemia
  - Thyroid function tests
  - Renal profile
  - Diabetes
  - Liver profile, coagulation (right heart failure)
- Coronary angiography
- ? Viral screen, family evaluation etc
Management – systolic impairment

- **Acute management – stable patient**
  - Oxygen
  - Diuretic therapy (loop +/- thiazide)
  - Treat any treatable causes (ischaemia, TFTs anaemia)
  - ACE inhibitor
  - +/- morphine and nitrate

- **Acute management – unstable patient**
  - Positive airway pressure oxygen
  - Diuretic
  - +/- morphine and nitrate
  - Consider intubation and ventilation
Long-term management – systolic impairment

- **Symptom relief**
  - Diuretics – loop +/- thiazide
  - Digoxin – even in sinus rhythm

- **Prognostic benefit**
  - ACE inhibitors (uptitrate slowly)
  - Beta-blockers (start after diuretics reduced or withdrawn, uptitrate gradually)
  - Aldosterone antagonist
  - Angiotensin receptor blockers (meta-analysis suggests not in combination with ACE i)
  - Hydralazine and nitrates
Long-term management – other issues

- **Diastolic dysfunction**
  - Symptomatic improvement with diuretic therapy
  - Caution not to over-diurese
  - No drug therapy of proven prognostic benefit

- **Right heart failure**
  - Diuretic therapy - spironolactone
  - Remove exacerbating elements (eg OSA)
  - Pleural / ascitic tap
Non-pharmacological Rx

- Exercise
- Restrict salt intake
- ? Reduce fluid intake (controversial)
- Reduce excessive weight
- Heart failure clinics
Invasive therapy

- Biventricular pacemaker (CRT)
  - Broad QRS
  - NYHA Class III or IV (? Less now)
  - ‘dyssynchrony’ echo
- ICD – often with above
  - EF < 30-35% after 4 (or 12) weeks optimal therapy
- LVAD
- Cardiac Transplantation
Cardiac resynchronisation therapy
Inherited cardiomyopathy

Sarcomere mutation

Nuclear envelope
Cytoskeleton
Sarcomere

Cell adhesion gene
<table>
<thead>
<tr>
<th>Condition</th>
<th>Estimated no affected in Ireland</th>
</tr>
</thead>
<tbody>
<tr>
<td>HCM</td>
<td>1:500</td>
</tr>
<tr>
<td>ARVC</td>
<td>1:1,000 - 10,000</td>
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<tr>
<td>DCM</td>
<td>1:3,000 - 5,000</td>
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</tbody>
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HCM - under a microscope
Genetic causes
Genetics of HCM

- 90% cases are Autosomal Dominant
  - Autosomal = not on X or Y chromosomes
  - Dominant = only one ‘abnormal’ copy of gene required to cause condition
  - 50% chance of affected person passing on ‘abnormal’ gene to each of their children
- Incomplete penetrance
  - Not everyone who inherits gene will develop symptoms and signs of the condition
- 10% ‘Sporadic’ – ie no family history of HCM
How do people present

- **Symptoms:**
  - Chest pain
  - Shortness of breath
  - Palpitations
  - Pre-syncope / syncope
  - Cardiac arrest

- **No symptoms:**
  - Incidental finding
  - Family screening
How is HCM diagnosed

- History
- Physical exam (‘jerky’ pulse, systolic murmur left sternal edge)
- ECG (LVH +/- ST segment and T-wave changes)
- Transthoracic Echo
- +/- Cardiopulmonary exercise test
- +/- MRI
- +/- Genetic testing
Complications of HCM

- None
- Chest pain
- LVOT obstruction
- Atrial fibrillation
- Thrombo-embolic events (strokes)
- Heart failure
- Cardiac arrest
Management of HCM

- **Symptom control**
  - B-blockers
  - Verapamil
  - Negative inotropes (LVOT obstruction)
  - Heart failure therapy
  - Anticoagulation
  - Septal reduction therapy (alcohol septal ablation vs surgical myectomy)
  - Transplant
HCM Management

- Genetic aspects
  - First degree relatives evaluated with ECG and Echo
    - 12-18 year-old annually
    - Thereafter 2-5 yearly
  - Genetic testing
- Sudden death risk
Magnitude of Sudden Cardiac Death in HCM

Percentage SCD in HCM

Causes of SCD in the Young

1.4%
Who is at risk?

- **Features that may indicate increased risk:**
  - Previous cardiac arrest
  - Family history premature sudden death
  - Unexplained blackouts
  - Abnormal rhythm on exercise test or Holter monitor (even 3 beats of ventricular tachycardia)
  - Blood pressure fails to rise normally with exercise
  - Severe thickening of the heart (>3 cms or almost 3x normal)

- Some more significant at younger age
- Presence of LVOTO may also increase risk
- Presence of fibrosis as indicated by late enhancement post contrast on cardiac MRI
Inherited DCM

- 7-25% of ‘idiopathic’ DCM may be familial
- Sometimes associated with more generalised myopathies (e.g., muscular dystrophies)
- Genetic causes may exceed 100 genes
- Presence of conduction disease may suggest specific subtypes
- Management same as for heart failure
Arrhythmogenic right ventricular cardiomyopathy (ARVC)

- Commonest cause of SCD in athletes in Northern Italy.
- Prevalence 1:1000 (or 1 in 5000).
- Familial in 30-40% of cases (or > 90%).
- Fibrofatty replacement of the right ventricle.
- Fatal ventricular arrhythmias of right ventricular origin.
Treatment of ARVC

- Asymptomatic, normal pump function, no abnormal rhythms detected: No Treatment
- Palpitations: Beta-blockers or Amiodarone
- Reduced pump function: ACE inhibitors, Spironolactone, diuretics etc.
- High risk: Implantable defibrillator
- End-stage pump failure: transplant
Sudden Cardiac Death

- Sudden Cardiac Death = death from definite or probable cardiac causes within 1 hour of symptom onset

- Incidence from International Studies
  - 1 to 3 per 100,000 in those 1 to 35 yrs of age
  - 10 to 75 per 100,000 in those 35 to 64 yrs

- Incidence in Ireland unknown

- Extrapolation from other studies suggest
  - > 5,000 SCD annually RoI, >2000 NI
  - > 60 deaths < 35 yrs (RoI), >25 (NI)
Causes of SCD

- Over 35 yrs of age, Coronary Heart Disease is most common cause
- Under 35 yrs
  - Cardiomyopathies
  - Congenital Heart Disease
  - ‘Structurally Normal Heart’ (ion channel disorders, conduction disease)
  - Anomalous coronaries
Sudden Adult Death Syndrome

- Cause not apparent on PM
- Cave potentially spurious causes
  - Non-obstructive coronary disease with no infarct
  - ‘LVH’ with normal heart weight
  - ‘sudden death in epilepsy’
- 40% of families have inherited cause identified (mostly LQT and Brugada)
Long QT syndrome

- Inherited form 1 in 5000
- Up to 7 genes associated
- Up to 75% have LQT1 (35-40%), 2 (30-35) or 3 (10%)
- Patients present with syncope or sudden death due to polymorphic ventricular tachycardia (torsades de pointes)
- LQT1 events occur with exercise or emotion
  - swimming
- LQT2 events occur with ‘startle’
- LQT3 events occur at rest or during sleep
Management LQT

- Avoid precipitants
  - Medications
  - Stimulants
  - Exercise (LQT types 1 and 2)

- B-blockers
  - LQT type 1 and 2

- ICD
Prevalence unknown

May be significant regional variation

Association of incomplete RBBB in right precordial leads with ST segment elevation and sudden death

‘Concealed’ cases may be unmasked by provocation tests

Management

- At risk if syncope or spontaneously abnormal ECG
- ICD currently only available treatment
Other causes SCD

- Myocarditis
- Catecholaminergic Polymorphic VT
- Congenital heart disease
- Questionable causes
  - Anomalous coronaries
  - MVP
  - Sudden death in epilepsy
Summary

- Heart failure widespread
- Prognosis still poor, but intensive management improves
- SCD in the young is rare but causes under-recognised
- Specialist centre for evaluation of and management of those at risk in Tallaght
  - Made possible by your hard work!!