

Disorders of the Myocardium – Cardiomyopathy, Myocarditis, ASD, VSD

Sept 2014

HOCM

50% familial with autosomal dominant inheritance
Systolic murmur decr with passive leg raising
Syncope worrying, precedes sudden cardiac death
Avoid vigorous exercise

ECG Changes

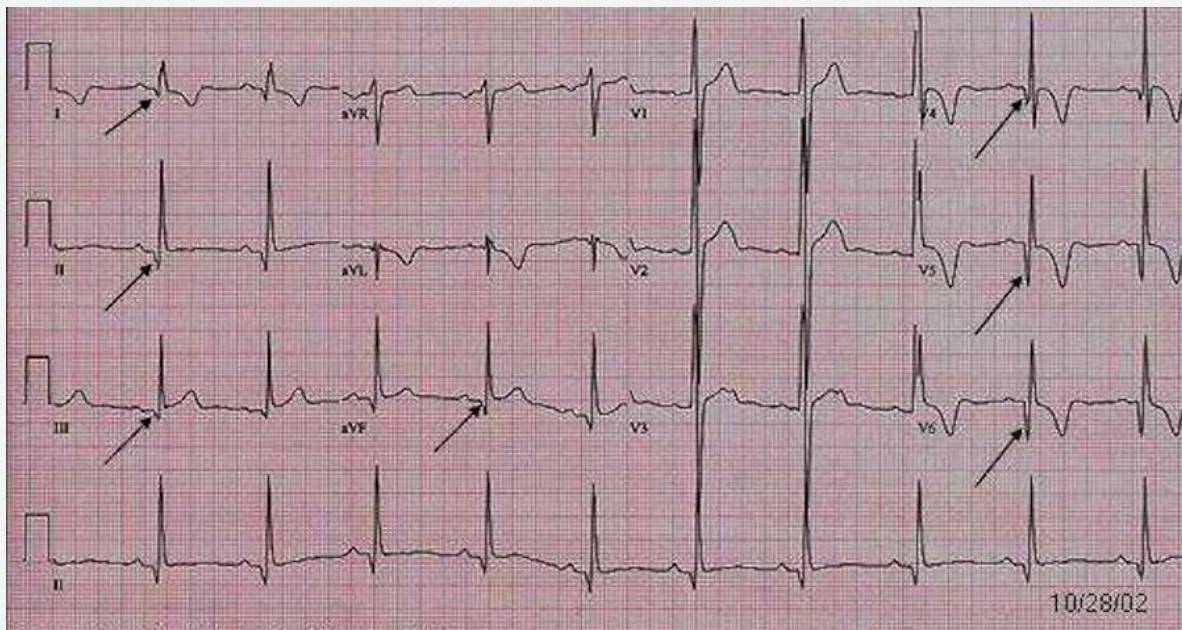
Classic: septal Q waves = large Qs in anterior, inferior or lateral

Plus:

1. Nonspecific ST/T changes - T inversions, large Ts
2. LVH +/- LAA (left atrial abnormality)
3. Atrial arrhythmias - AF (poorly tolerated, decr filling)
4. PACs, PVCs

NB Septal Q wave: "pseudoinfarction" - ie looks like infarct

How to differentiate: Upright T suggests pseudoinfarction; Inverted T suggests infarction



Restrictive Cardiomyopathy

Most common causes: amyloidosis, scleroderma, carcinoid, sarcoidosis

Dilated Cardiomyopathy (DCM)

Ventricular dilatation and global myocardial dysfunction (ejection fraction < 40%)

Symptoms of biventricular failure, e.g. fatigue, dyspnoea, orthopnoea, ankle oedema.

High mortality (2-year survival 50%) due to progressive cardiogenic shock (sudden cardiac death)

Causes

Ischaemic - post massive anterior MI

Non-ischaemic - most idiopathic, 25% familial, some due to viral myocarditis, alcoholism, toxins (doxorubicin), autoimmune, pregnancy (peripartum cardiomyopathy)



ECG Changes

No unique ECG features, although ECG usually abnormal
Most common: left or bi-atrial or ventricular hypertrophy
LBBB due to cardiac dilatation
Reduced voltages due to myocardial fibrosis
Abnormal Q waves V1 to V4 - "pseudoinfarction" pattern
AF

Myocarditis

More common in children and young adults; common cause of sudden death in previously well

Causes

Viral
Autoimmune
Bacterial (Q fever, N meningitidis, M pneumoniae, C diphtheriae, chlamydia, beta haem strep)
Parasitic (Chagas disease most common cause worldwide, toxoplasma)
Drugs (doxorubicin, ETOH, clozapine, radiation)

Symptoms

Non-specific; weakness, SOB, CP, fever, dizziness, palpitations, recent flu-like illness
Incr HR out of proportion to fever, incr RR, hepatomegaly, creps, normal/high BP but poor perfusion, maybe rapid deterioration, LVF uncommon

Investigations

ECG - Tachycardia, low ECG voltages in 80%, ST/T changes, conduction disturbances, long QTc
CXR - Cardiomegaly, pleural effusions
ECHO - global decr contractility, decr EF, V dilation
Incr cardiac markers; incr ESR in autoimmune
Myocardial biopsy (50-70% sens)

Management

Supportive - CCF treatment; bed rest; inotropes, diuretics, vasodilators, ACEi, Treat arrhythmia
Mechanical support if hypoperfusion despite meds (ECMO, V assist devices)
Steroids/immunosupp if autoimmune

Prognosis

Most deaths in 3-4/7; >95% survival if last >72hrs
Often progresses to DCM
No competitive sport for 6/12

ASD

Haemodynamically significant if > 2 cm diameter
Ostium Secundum most common = 75%, but least serious
Ostium Primum 2nd most common, more symptomatic
Childhood heart failure & incr mortality without surgery
Most pts asymptomatic until adolescence - pulmonary HTN, arrhythmias, heart failure develops



Examination

Generally well perfused patients
Fixed split 2nd heart sound
Pulmonary systolic murmur from increased flow over normal pulm valve
Mid diastolic flow murmur if large defect
ASD itself = no murmur

Investigations

CXR - Cardiomegaly, Prominent right atrium and pulmonary artery
ECG - AF common after 30-40 years, RAD, RV hypertrophy, P pulmonale, prolonged PR, RBBB
Echo

VSD

Most common cardiac defect
60% close in late childhood
70 % located in membranous portion of intra-ventricular septum
25 % have other cardiac abnormality

2 types

Perimembranous VSD (75%) - opening in upper section of ventricular septum, near valves
Muscular VSD - opening in lower section of ventricular septum

Clinical presentation

Symptoms & signs depend on defect size, presence/absence of other defects
Increased risk for Infective Endocarditis
Small defect - Often found on clinical exam, > 60% close spontaneously in later childhood
Moderate defect - Cause increased RV pressure = pulmonary HTN
Infants with increased cough with mild urti, may have mild incr pulmonary vascularity/early CHF
Large defects - Cause CHF early in infancy = early and severe increase in pulmonary artery pressure = pulmonary HTN
if uncorrected = progress to Eisenmenger syndrome (reverse of L-R shunt with cyanosis)

Fatigue, sweating, tiring when feeding, poor weight gain, tachypnoea, tachycardia
Clubbing, may be cyanosed
Dynamic left ventricular impulse & weak right ventricular impulse
Pan systolic murmur, loudest at LSB & radiates to back (Smaller VSD = louder murmur)
So loud murmur in child who is feeding well = probably a small VSD
Pulmonary systolic murmur with increasing severity
Mitral diastolic murmur

Investigations

CXR - Cardiomegaly & Biventricular enlargement and Pulmonary blood flow; ECG, Echo

Complications

Left to right shunt; Eventually reversal of shunt (becomes Right - Left) = Eisenmengers
Pulmonary Hypertension; RVF/Hypertrophy
Risk endocarditis

Management

Small ones close spontaneously, larger ones need a patch
Medical: digoxin, diuretics, nutritional supplements, endocarditis prophylaxis