Overview of Cardiomyopathy

1. Describe the conditions under this heading

Overview of Cardiomyopathy

Cardiomyopathy typically meant cardiac dysfunction without one of the well-known causes of cardiac failure being present:

* Infarction
* Valvular disease
* Myocarditis
* Cor pulmonale

The disease was then defined by its anatomical picture:

|  |  |  |
| --- | --- | --- |
|  | Anatomical picture | Non-cardiomyopathy causes |
| Dilated | Thin-walled, dilated myocardium | Common consequence of ischaemic. |
| Hypertrophic | Irregularly thickened myocardium | Classically seen in:   * Hypertension * Aortic stenosis |
| Restrictive | Thickened, non-dilating ventricles | May be part of heart failure in any other pattern. |
| Other | Various:   * Obliterative cardiomyopathy * Restrictive cardiomyopathy * Takotsubo cardiomyopathy | These forms have unique features which distinguish them from other forms. |

These forms are now understood as diseases in their own right with well-defined characteristics but the old terminology lingers.

# Genetic disease

Two forms are now understood to be strictly genetic conditions:

1. Hypertrophic cardiomyopathy  
   *Inappropriate, irregular and elaborate hypertrophy of the LV*
2. Arrhthmogenic ventricular cardiomyopathy  
   *Irregular patches of fibrous/fatty tissue appear in the myocardium (usually the RV)*

Both forms are inherited in an **autosomal dominant** fashion, and associate with a heritable risk of sudden cardiac death.

# Secondary disease

Three forms are typically seen in association with a systemic disease:

1. Dilated cardiomyopathy  
   *Wide variety of associations: alcohol, post-myocarditis, multiple genetic forms*
2. Restrictive cardiomyopathy  
   *Usually seen with amyloidosis or, rarely, glycogen storage diseases*
3. Obliterative cardiomyopathy  
   *Associated with eosinophilic disease (leukaemia, Churg-Strauss)*

## Other causes

Clearly many other things can cause cardiac damage, however these perhaps do not identify with such clearly defined diseases as the classic cardiomyopathies.

# Spontaneous disease

Takotsubo cardiomyopathy occurs without other physical disease, although environmental or emotional stressors are often present.

It has a characteristic presentation:

* Chest pain, dyspnoea, and cardiac failure
* ST elevation on ECG
* LV aneurysm on echo

Clearly this must be carefully distinguished from acute ischaemia, but the prognosis is excellent.