

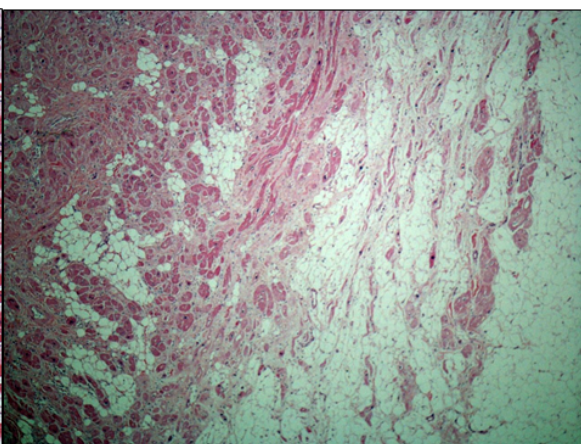
Table 1. The diagnostic criteria that Mary N Sheppard has developed and uses to assess for the major cardiac diseases that underlie SCD.

Pathology	Macroscopic Criteria	Microscopic Criteria
Hypertrophic cardiomyopathy	Increase in heart weight*. Right ventricular wall thickness >5mm or left >15 mm. Atrial dilatation. Can be normal macroscopically. Absence of coronary artery disease.	Left ventricular myocyte disarray (>20% of myocardial disarray in at least 2 cardiac sections) and myocyte hypertrophy with or without interstitial or replacement fibrosis and thick walled blood vessels.
Arrhythmogenic cardiomyopathy	Normal or increased heart weight*. Right or left ventricular thinning, fatty replacement, fibrosis on the epicardial surface. Can be normal macroscopically. Absence of coronary artery disease.	Fibrosis admixed with fatty infiltration of the myocardium originating from the epicardial surface (>20% in at least 2 cardiac sections).
Dilated cardiomyopathy	Increase in heart weight* with dilated left ventricle (>40mm chamber diameter) and thin compact wall (<10mm). Mural thrombi in ventricles Dilated atria with thrombi in appendages. Absence of coronary artery disease.	Widespread diffuse interstitial or replacement fibrosis (>20% in at least 2 cardiac sections) in the left ventricle with atrophic myocytes.
Idiopathic left ventricular hypertrophy	Increase in heart weight*. Left ventricular wall thickness >15 mm. No hypertension or coronary artery disease.	Myocyte hypertrophy with or without replacement or interstitial fibrosis. Absence of myocyte disarray.
Idiopathic left ventricular fibrosis	Normal heart weight and wall thickness with/without scarring macroscopically. Absence of coronary artery disease. Absence of coronary artery disease.	Replacement or interstitial fibrosis (>20% in at least 2 cardiac sections). Absence of myocyte disarray.
Hypertensive heart disease	Increase in heart weight*. Left ventricular wall thickness >15 mm. History of hypertension.	Myocyte hypertrophy with fine interstitial fibrosis in subendocardium. Absence of myocyte disarray.

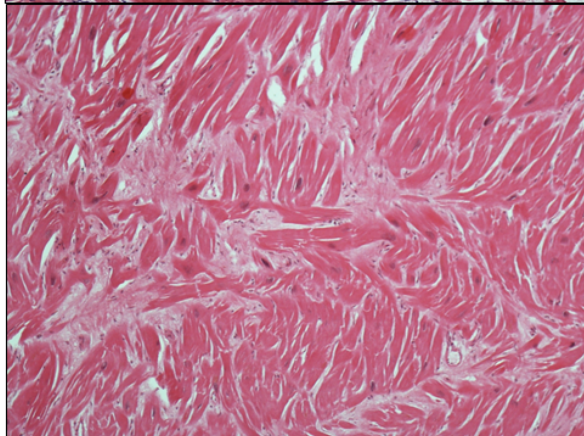
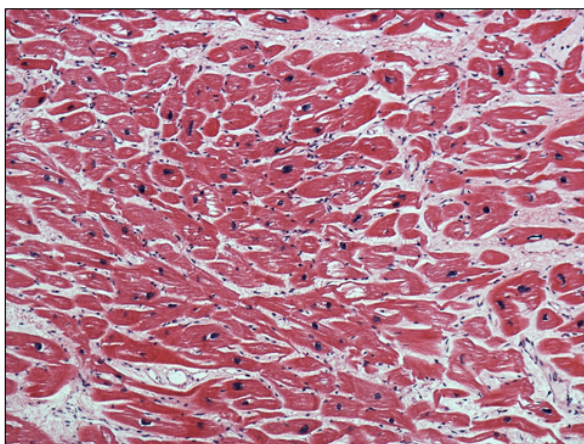
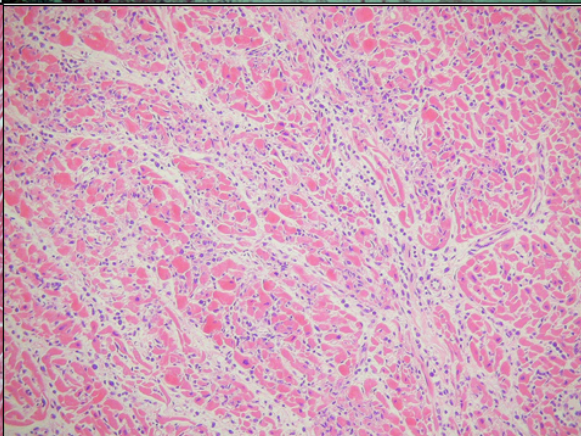
	No coronary artery disease.	
Myocarditis	Normal or dilated ventricles with variegated appearance. Fibrinous pericarditis. Can be normal macroscopically.	Inflammation (>20% in at least 2 cardiac sections) with associated myocyte necrosis.
Coronary atherosclerosis	Atherosclerosis with luminal narrowing >75% or lumen less than 1mm or inability to insert 2mm probe. Infarction or scarring in myocardium. Normal myocardium Rupture with haemopericardium. Thrombosis in coronary artery.	Acute or chronic infarction in right or left ventricle. May be no infarction.
Anomalous coronary artery	Anomalous origin of the coronary artery in the incorrect sinus with interarterial course or pulmonary artery origin.	May have acute or chronic infarction in the right or left ventricle.
Mitral valve prolapse	Prolapse of mitral valve above the atrioventricular junction with ballooning between cords in one or both leaflets. Diffuse thickening of leaflets. Mitral Annular dilatation. Cordal thinning and /or rupture.	Myxoid degeneration with expansion in spongiosa of leaflets and destruction of fibrosa layer. May be subendocardial fibrosis in posterobasal left ventricle.
Aortic stenosis	Significant valve stenosis demonstrated by inability to insert a finger through the annulus. Calcified valve and/or bicuspid valve. Increase in heart weight* and left ventricular wall thickness >15mm.	Myocyte hypertrophy and/or interstitial or replacement fibrosis throughout left ventricle.

Footnotes: *Increase in heart weight is defined as greater than 450g in females and greater than 550g in males.

ACM



Myocarditis



ILVH

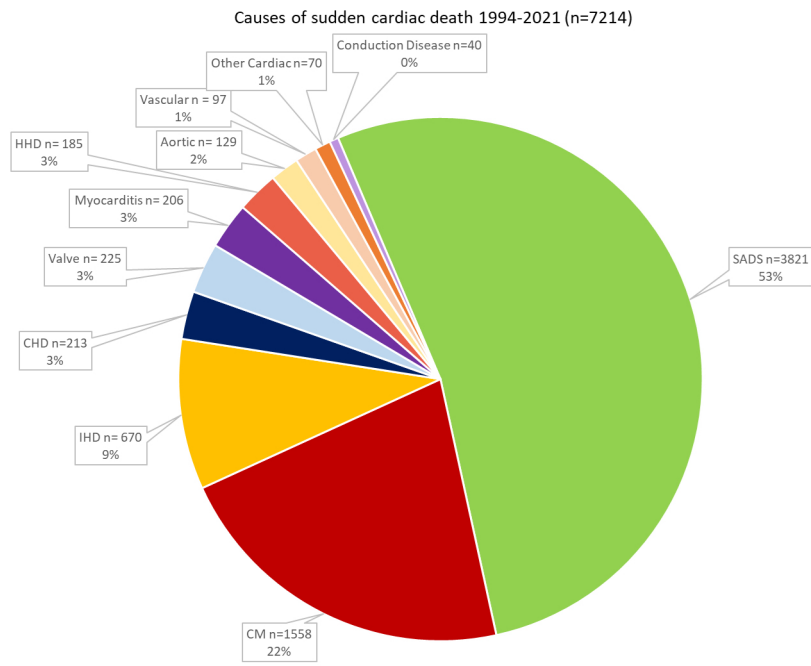
Figure legend:

Upper left: Idiopathic left ventricular hypertrophy showing hypertrophied myocytes.

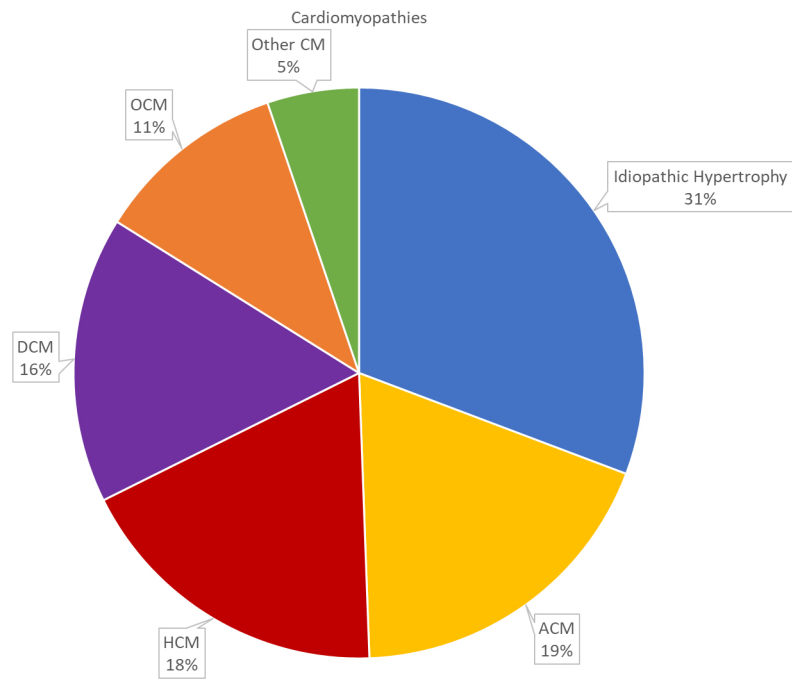
Upper right: Arrhythmogenic cardiomyopathy showing fat and fibrosis present in the subepicardium.

Lower left: Hypertrophic cardiomyopathy showing myocyte disarray.

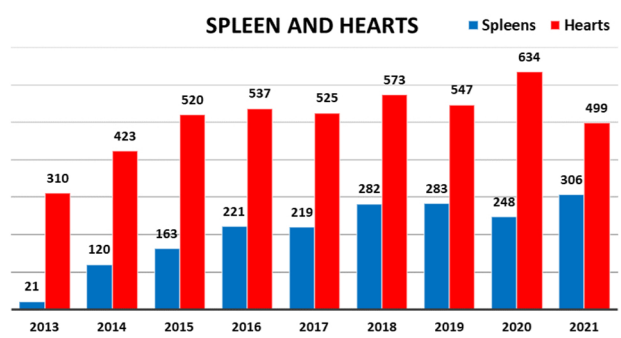
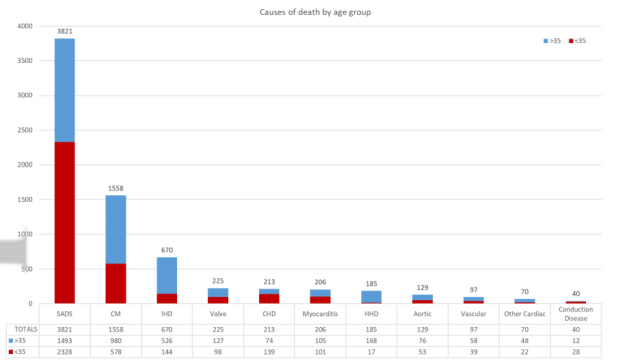
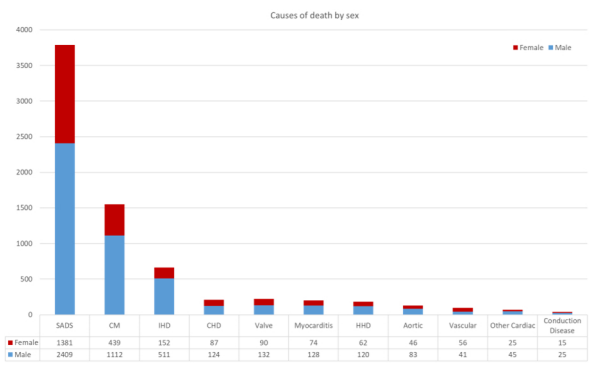
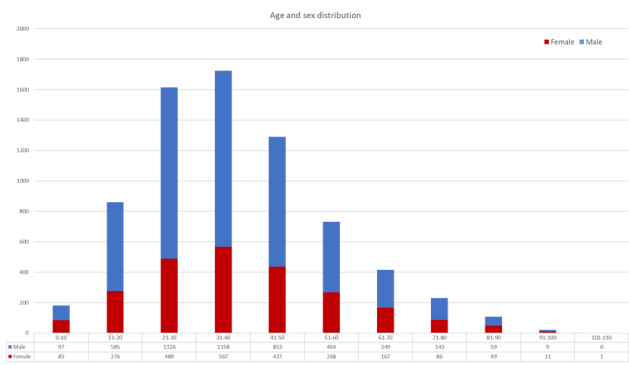
Lower right: Myocarditis showing a lymphohistiocytic inflammatory infiltrate with corresponding myocyte necrosis.



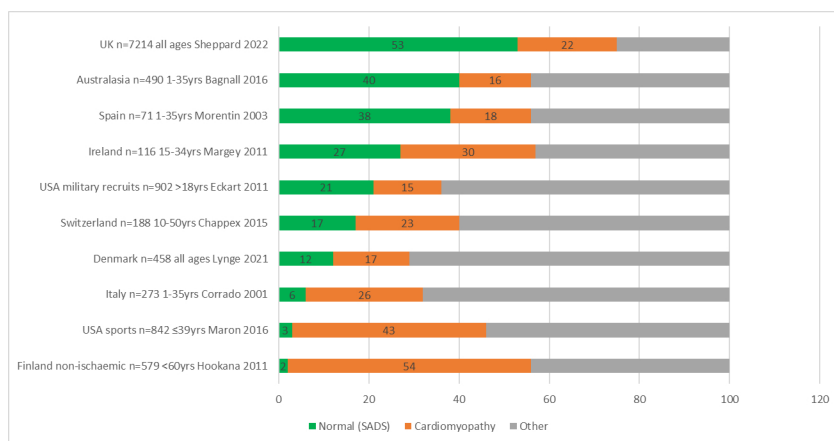
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HIS_14889_Cardiomyopathy pie.jpg



HIS_14889_Graph panels.jpg



HIS_14889_SADS CM proportions reported worldwide.jpg