The paper of the month: *The Classification Concept of the ESC Working Group of Myocardial and Pericardial Diseases*

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The term *cardiomyopathy* was first used over forty years ago, to describe myocardial disorders that are not caused by haemodynamic disturbances such as valve disease and hypertension, or by multi-system diseases. Heart muscle disorders with an identifiable aetiology were initially termed specific heart muscle diseases, but were later renamed specific cardiomyopathies. Remarkably, this nomenclature has survived to the present day with only minor changes. Recently, expert committees of the AHA and the ESC Working Group on Myocardial and Pericardial Diseases have proposed updates of the cardiomyopathy classification system [1,2]. The motivation of both groups was to resolve outstanding ambiguities in the existing classification and to incorporate knowledge derived from advances in molecular genetics. Both proposals continue to define cardiomyopathies according to the morphology and physiology of the ventricles and emphasize the importance of genetic disease as a cause of cardiomyopathy. The major differences relate to the handling of primary and secondary disease and the classification of ion channel disorders

Summary

A clinical classification should be simple and relevant to everyday medical practice. The ESC working group's classification achieves this goal, and by highlighting the importance of familial disease, will result in more accurate diagnoses and better outcomes for patients.

The AHA classification

Cardiomyopathies have always been divided into primary and secondary forms. The definition of primary has, however, been somewhat ambiguous, interpreted by many to mean idiopathic and others as disease confined to the heart (i.e. not the consequence of a systemic disorder). The new AHA proposal maintains this taxonomy, defining primary cardiomyopathies as diseases solely or predominantly confined to heart muscle and secondary cardiomyopathies as cardiac disorders with "pathological myocardial involvement" as part of generalized systemic disorders". The first major departure from the existing convention is the sub-classification of primary cardiomyopathies into genetic, mixed (genetic and non-genetic) and acquired forms. The second, and much more controversial change, is the redefinition of ion channelopathies as primary, genetic cardiomyopathies.

The ESC Classification

The ESC working group's proposal abandons the distinction between primary and secondary altogether. The existing morphologic sub-types of cardiomyopathy (hypertrophic, dilated, restrictive, *etc*.) are retained with some modifications, but they are separated into familial (or genetic) and non-familial (or non-genetic) sub-types. These are then further subdivided into known genetic causes, idiopathic and acquired diseases as appropriate. The principle aim of this simplification of the old classification was the encouragement of a shift from the existing exclusion-based diagnostic paradigm towards a more logical and thorough search for diagnostic markers.

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Comparison of the AHA and ESC systems

Both the ESC and AHA panels recognized that the current classification system has some important limitations. They also have a common purpose in seeking to assist diagnostic and therapeutic decision-making. The AHA and ESC systems use similar morphological and physiological criteria to describe sub-types of cardiomyopathy and both sub-classify disease into genetic and non-genetic forms. Of the differences between the two systems, perhaps the least important is the primary versus secondary issue. The ESC working group's view was that this distinction has always been arbitrary and that abandonment of the terminology results in a simpler and more consistent approach to classification of heart muscle disease. The classification of ion channellopathies as cardiomyopathies by the AHA panel is quite another matter. The ESC working group's view is that it is inappropriate to reclassify a whole group of diseases that have little or no clinically detectable effect on cardiac morphology and haemodynamics. The AHA panel's decision to do so is predicated on the idea that mutations in ion channel genes result in changes in protein structure within cardiomyocytes and therefore can be regarded as disorders of heart structure. They also cite reports of dilated cardiomyopathy caused by mutations in the sodium channel gene. The problem for the ESC working group is that the majority of cardiologists use the term ion channelopathies as shorthand for a group of syndromes defined *entirely* by specific electrocardiographic criteria, often excluding patients that have evidence for structural heart disease. For this reason, the ESC working group felt that it is premature to create a whole new class of arrhythmogenic cardiomyopathies. If it transpires that mutations in ion channel genes are an important cause of structural heart disease, the new ESC classification is flexible enough to acknowledge them as one of the many causes of existing cardiomyopathy subtypes.

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References

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2. *Elliott P, Andersson B, Arbustini E, Bilinska Z, Cecchi F, Charron P, Dubourg O, Kühl U, Maisch B, McKenna WJ, Monserrat L, Pankuweit S, Rapezzi C, Seferovic P, Tavazzi L, Keren A. Classification of the cardiomyopathies: a position statement from the european society of cardiology working group on myocardial and pericardial diseases. Eur Heart J. 2008;29(2):270-6.*