This paper was the first unequivocal morphological description of the septal form of hypertrophic cardiomyopathy in nine subjects dying suddenly at a young age. It highlighted sudden death as an important feature of the disease and provided the probable morphological basis for other patients found to have left ventricular outflow obstruction at a subaortic level in life. [The SCI® indicates that this paper has been cited in over 390 publications, making it the most-cited paper from this journal.]

First Demonstration of Septal Hypertrophic Cardiomyopathy

M.J. Davies
Cardiovascular Pathology Unit
St. George's Hospital Medical School
London SW17 ORE England

August 12, 1989

Donald Teare's paper is a Citation Classic in providing the first unequivocal illustration of the septal form of what is now known as hypertrophic cardiomyopathy (HCM). Teare's paper caught the clinician's eye because his method of dissecting the heart, somewhat unorthodox by manuals of pathologic dissection, illustrated that the septum bulged out into the left ventricular outflow tract. Clinical cardiologists had been beginning to recognize subaortic stenosis in living subjects, and it was not difficult to appreciate that the morphological appearances described by Teare were the likely substrate. The paper also was striking in that nine cases of sudden death in young subjects were described from six families; this highlighted two of the clinical features, a familial trend and high risk of sudden death, that characterize HCM.

Teare, with whom I became associated in 1964 up to his death in 1979, did not have a special interest in hearts. He was a forensic pathologist carrying out many hundreds of autopsies per year on victims of sudden natural death to supplement the rather modest salary paid for true forensic work. He did however have the innate ability to recognize morphological abnormalities and the intellectual curiosity to store hearts and other organs away for future consideration. The concordance of a similar abnormal morphological appearance of the hearts in two subjects from one family dying suddenly at a young age prompted a search for other hearts with a similar appearance in his collection. As a consequence the paper concentrates solely on the septal form of HCM, and indeed had not clinical interest also been centered on this form of the disease, the paper might have gone unnoticed.

Teare described accurately the characteristic histological appearances in the myocardium of muscle bundles running in diverse directions and separated by connective tissue giving "an impression of inefficiency in muscular contraction." These appearances would now be called myocardial disarray and are regarded as specific on a quantitative basis for the disease. Teare interpreted the histology as indicating a benign tumour or hamartoma of the heart.

The acceptance of a histological "gold standard" in the form of myocardial disarray, albeit only really useful at autopsy with the whole heart available, and the growth of echocardiography has led to the realization that asymmetric septal hypertrophy is by no means the only morphological expression of HCM. Teare himself realized there was a far wider spectrum to HCM. In 1974 he and I wrote a paper describing symmetric involvement of the left ventricle and highlighted the fact that although sudden death in young subjects is a common presenting feature, cases with identical morphology were found in elderly subjects at necropsy. HCM is now known to be a process that can involve any portion, or indeed the whole of the left ventricle, as well as involving the right ventricle. In parallel with the heterogeneous morphological expression is a wide range of clinical manifestations. It is now accepted that hypertrophy in the affected segment of myocardium develops in the first two decades of life, and there is no indication from echocardiographic studies of spread to involve the rest of the ventricle. As a corollary there will be cases with the disease in children or adolescents who have not yet developed their full morphological expression of the disease.

The question remains whether there is one disease with a very variable morphological and clinical expression, but a common histological basis, or several diseases that share a common histological appearance. The identification of the gene in families with "typical" asymmetric septal HCM is awaited.