

Editorial

Hypertrophic cardiomyopathy in the elderly

Hypertrophic cardiomyopathy, defined as idiopathic hypertrophy of a non-dilated ventricle, has a range of morphological and clinical features and can occur from infancy to old age.¹ Hypertrophic cardiomyopathy, although initially regarded as a rare disorder of young patients with symptoms, is probably not uncommon. In the early years, after the first descriptions of the disease, most clinical studies came from a few tertiary referral centres²—this biased selection towards young patients with symptoms who had a disease characterised by asymmetrical septal hypertrophy and dynamic left ventricular outflow tract obstruction.³⁻⁵ Since then, however, it has become apparent, particularly with the widespread application of echocardiography, that hypertrophic cardiomyopathy occurs in patients of all ages with or without regional septal hypertrophy, an outflow tract gradient, or symptoms.⁶⁻¹²

Diagnosis

Echocardiography is now the technique of choice for the diagnosis and assessment of hypertrophic cardiomyopathy. The echocardiogram usually shows hypertrophy of the ventricular septum, but any region of the left ventricle (and occasionally the right ventricle) can be affected. Left ventricular contraction is hyperdynamic and there is often impaired and incoordinate relaxation and a reduced rate of filling.¹³⁻¹⁶ These features are similar in all age groups, but in the elderly fibrous thickening of the ventricular septum in the area of mitral septal contact may be prominent and there can be quite extensive mitral annular calcification.¹⁷ The aortic valve may also be calcified and regurgitant.

In a young patient, the echocardiographic finding of idiopathic left ventricular hypertrophy may lead with certainty to the diagnosis of hypertrophic cardiomyopathy; but in the elderly there are more frequent secondary causes of left ventricular hypertrophy. In particular, increasing age is clearly associated with increased left ventricular mass and septal thickness and changes in diastolic function.¹⁸⁻²² In extreme old age, the septum may not only be hypertrophied but also sigmoid, which can also simulate the appearance of hypertrophic cardiomyopathy.²³ In children with clinical evidence or a family history of hypertrophic cardiomyopathy rapid development of left ventricular hypertrophy can be seen by echocardiography over four years. Increases in wall thickness of up to 23 mm have been noted.²⁴ In contrast, a similar study in 65 adults showed that none demonstrated an increase in left ventricular wall thickness and 14% had a significant reduction with relative enlargement of the left ventricular cavity.²⁵ In addition, echocardiographic measurements of left ventricular mass and maximum wall thickness were significantly higher in patients aged 21-30 than in older patients. Younger patients also had the most severe and diffuse hypertrophy in a study of 700 individuals with hypertrophic cardiomyopathy.²⁶ However, some patients, particularly women, may present in their sixth or seventh decade after a prolonged asymptomatic (latent?) period with progressive symptoms and often a poor response to medical treatment.

Such patients may have a distinctive type of the disease, with modest septal hypertrophy, distortion of the left ventricular outflow tract, and a gradient generated by the posterior motion of the septum against the restricted movement of a calcified mitral valve.¹⁷

Prognosis

While premature sudden death is a well-known feature of hypertrophic cardiomyopathy it is relatively uncommon in patients with hypertrophic cardiomyopathy.^{27,28} Sudden death is not confined to the young but most studies of the natural course of the disease have emphasised its rarity in elderly patients.^{29,30} A retrospective analysis of 284 patients showed that sudden death was more common in those aged < 14 years and in older patients with symptoms. The annual mortality for cases diagnosed in childhood was 5.9%, compared with 2.5% for cases in adults.³¹ The better prognosis of elderly patients may reflect the survival of a subset of patients at low risk of sudden death, especially those with ventricular tachycardia or an adverse family history.^{32,33} Lack of exertion in the elderly may also have a protective effect.

Hypertensive hypertrophic cardiomyopathy?

In 1957, when Brock first described an elderly patient with severe left ventricular hypertrophy and an outflow tract gradient, he concluded that the disease was secondary to hypertension³⁴; this was not borne out in subsequent reports.^{35,36} Though the morphological and echocardiographic features of hypertrophic cardiomyopathy and severe secondary left ventricular hypertrophy are usually distinctive,³⁷ diagnosis is complicated in mildly hypertensive patients with unequivocal echocardiographic features of hypertrophic cardiomyopathy in whom the degree of hypertrophy is inappropriate for the severity of hypertension. This problem was emphasised by the identification of 21, mostly elderly, female patients with severe concentric hypertrophy and small left ventricular cavity with hyperdynamic left ventricular contraction and abnormal diastolic function.³⁸ These patients usually presented in heart failure and their symptoms tended to be relieved by β blockade or calcium channel blocking drugs, rather than the traditional treatment of heart failure. This condition (hypertensive hypertrophic cardiomyopathy) suggests the coexistence of hypertrophic cardiomyopathy and hypertension. Among patients with hypertrophic cardiomyopathy those with hypertension tend to be older than those without. The clinical and echocardiographic features of hypertrophic cardiomyopathy were indistinguishable in those with hypertension and those without, except that hypertensive patients had thicker ventricular walls.^{39,40} The wide variability of expression of hypertrophic cardiomyopathy implies that other stimuli to the expression of hypertrophy such as pressure overload and neurohormonal factors may be important in genetically susceptible individuals. Further careful clinical and genetic

studies are required to clarify whether the definition of hypertrophic cardiomyopathy needs to be enlarged to include hypertension or whether hypertensive hypertrophic cardiomyopathy is a distinct clinical entity.

Treatment

Patients of all ages without symptoms or arrhythmias do not require treatment. Dyspnoea or angina are often adequately managed with β blockade or calcium antagonists^{41,42} and amiodarone improves prognosis in patients with arrhythmias.⁴³ Surgery should not be excluded in the elderly because the results of myotomy and myectomy in those with outflow tract gradients (with or without concomitant coronary bypass grafting) are good.^{44,45}

The presentation of an apparently congenital disorder in the elderly (without previous heart disease) is puzzling. While the elderly may present with typical symptoms of dizziness, syncope, breathlessness, chest pain, and palpitation the diagnosis is often delayed because these symptoms can be caused by coexisting disease. In addition, elderly patients can present in intractable heart failure caused by abnormalities of left ventricular relaxation and filling, and the diagnosis may be obscured by mitral regurgitation and mitral annular calcification. Although the distinctions between primary and secondary causes of hypertrophy are clear in young adults they are vague in the very young and the elderly. However, identification of the cause of hypertrophy probably has little influence on management.

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