

# Mitral Valve Endocarditis in Hypertrophic Cardiomyopathy

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## Citation

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## Abstract

Mitral valve endocarditis, as a complication of hypertrophic cardiomyopathy, usually affects the left ventricular aspect of the anterior mitral valve leaflet. It occurs predominantly in the presence of outflow tract obstruction. Even in the presence of hypertrophic cardiomyopathy with obstruction, the development of infective endocarditis is unusual. This article reviews the co-existence of these conditions and discusses treatment options, which include simultaneous mitral valve replacement and septal myectomy.

## ABBREVIATIONS

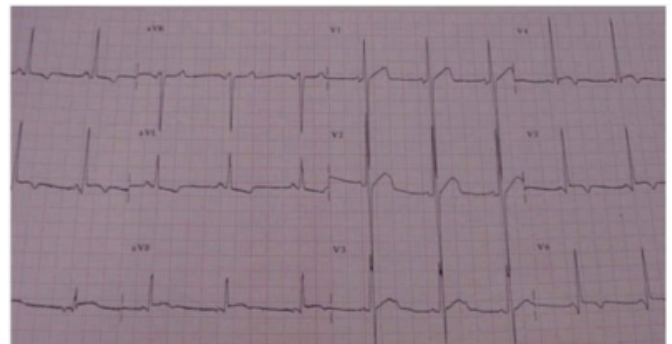
HCM, hypertrophic cardiomyopathy; OTO, outflow tract obstruction; IE, infective endocarditis

## INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a heterogeneous, hereditary condition of the cardiac muscle characterised by asymmetrical septal hypertrophy. This may result in left ventricular outflow tract obstruction (OTO), abnormalities of mitral valve motion (systolic anterior motion of the anterior mitral valve leaflet) and causes abnormalities of the electrocardiogram (Figure 1) and echocardiogram. Infective endocarditis (IE) is recognised as a complication of HCM and the 1997 American Heart Association guidelines recommend antibiotic prophylaxis in this disease. <sup>1</sup> More recently it has been demonstrated that the risk is virtually confined to patients who have OTO under basal conditions, and especially those with left atrial dilatation. <sup>2</sup>

## Figure 1

Figure 1: A typical electrocardiogram from a patient with hypertrophic cardiomyopathy, showing left ventricular hypertrophy and lateral repolarisation changes.



## PREVALENCE, INCIDENCE AND PATHOPHYSIOLOGY

In 1999, Spirito et al reported a study of 810 patients with HCM, which represents the most comprehensive study in the literature. <sup>2</sup> In the two decades prior to this only 33 patients with IE and HCM had been reported. Initial echocardiographic evaluation identified 3 cases of 'prior' IE, giving a prevalence of 3.7 per 1,000 patients. A further 2 patients had been referred actually because of the existence of IE and these were not therefore included in the analysis of incidence and prevalence. Subsequently a total of 681 patients were followed up for a mean of 55 months. During this time there were 5 patients who developed acute IE, giving an incidence of 1.4 per 1,000 person-years. In general therefore IE in the presence of HCM is rare with the cumulative 10-year probability as low as 5%.

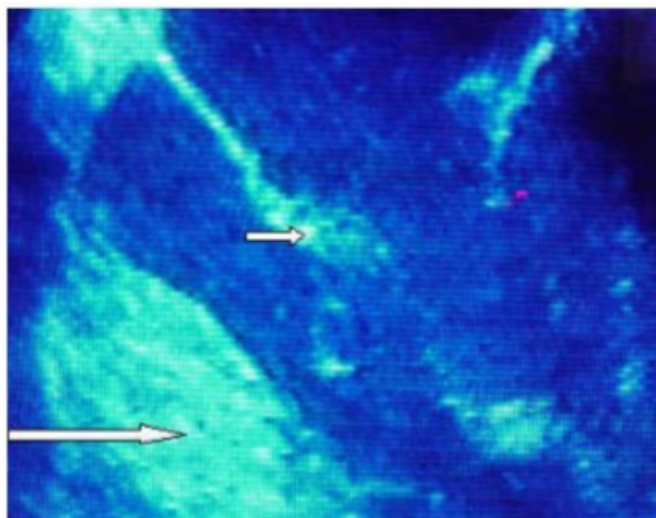
Studies examining the mitral valves from patients with HCM

and IE have found that the vegetations are most commonly situated on the left ventricular (or septal) aspect of the anterior mitral valve leaflet (Figure 2).<sup>3</sup> It has been presumed that this is due to the endothelial disruption caused by systolic anterior motion of the anterior mitral valve leaflet and resulting mitral-septal contact.<sup>3</sup> All of the 10 patients with IE in Spirito's series had vegetations on the mitral valve. Three patients also had involvement of the aortic valve.

Figures 2A and 2B: Transoesophageal echocardiography (4-chamber, mid-oesophageal, view) from a patient with hypertrophic cardiomyopathy and mitral valve endocarditis. Figure 2A shows marked septal hypertrophy (long arrow) and the anterior mitral valve leaflet, seen here in diastole, looks thickened (short arrow). There is a large vegetation attached to the left ventricular aspect of the anterior mitral valve leaflet, seen in Figure 2B (arrow).

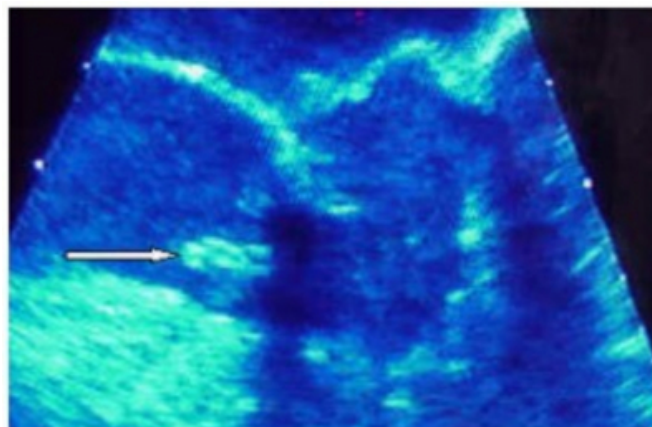
**Figure 2**

Figure 2A



**Figure 3**

Figure 2B



### CLINICAL FEATURES AND COURSE

Community acquired native valve IE in the general population is now most likely to be caused by staphylococci, usually *Staphylococcus aureus*, as it used to be caused by the oral ("viridans") streptococci. The presence or absence of HCM in IE patients has no bearing on the nature of the infecting organism. The organisms described in the literature on the subject reflect this, with Staphylococci and Streptococci predominant.<sup>2,4</sup> It is however noticeable from the recorded cases that the patients do tend to be younger (mean age in Spirito's series: 39 years) than is seen in general cardiology practice, outside of areas where rheumatic fever remains common.<sup>2</sup>

It is contentious whether the prognosis associated with IE is worse if there is underlying HCM. Some authors suggest that it is.<sup>5</sup> However the patients in Spirito's study actually did very well, with 8 of the 10 patients surviving and asymptomatic at follow up. Five of these patients had NYHA functional class III and IV heart failure symptoms at presentation. Of these 3 underwent mitral valve replacement (2 of whom also had aortic valve replacement), 1 underwent mitral valvuloplasty with associated septal myotomy-myectomy, and 1 showed clinical improvement on medical therapy alone.

### TREATMENT

The most efficacious treatment is of course prevention. However, despite the relationship between poor dentition and oral streptococcal IE, there is seldom a history of preceding dental treatment. The value of antibiotic prophylaxis in the prevention of IE has been questioned for some years, as there is little or no objective evidence that it is effective in preventing IE.<sup>6</sup> Although this scepticism

exists over the usefulness of antibiotic prophylaxis, guidelines (as has been alluded too) continue to support their use. It is clear from the literature that the potential benefit of this preventative measure only applies to those with the more severe cardiac structural abnormalities associated with HCM.

The mainstay of treatment in IE is antibiotic therapy, as guided by the organism cultured. <sup>7</sup> Definitive treatment for IE in HCM may be full medical therapy, or surgical intervention. Surgical treatment may be traditional (mitral valve replacement or even repair) but may also allow for simultaneous surgical treatment of the underlying condition, for example septal myectomy. Successful treatment even with *S aureus* as the infecting organism, has been described with medical therapy. <sup>4</sup> In general terms however, traditional indications, (haemodynamic, persistent fever, abscess, emboli) and relative indications (staphylococcus or fungal infection, renal impairment and vegetation size) for surgical intervention apply. <sup>8</sup> The combination of HCM with OTO and infective endocarditis should prompt early liaison with cardiothoracic surgical colleagues. If the clinical features suggest that mitral valve surgery is required, then the nature of the operation must be decided. There are two reported cases in the literature of mitral valve surgery combined with septal myectomy under these circumstances. <sup>9</sup>

Both mitral valve surgery and the Morrow septal myectomy are accepted methods of treating symptomatic obstructive HCM refractory to medical treatment, but the largest series on the surgical management of HCM suggests that the two operations are rarely combined. <sup>10 11 12 13</sup> Expertise in performing the Morrow procedure is found in selected centres, and not all cardiothoracic surgeons would necessarily be experienced in HCM surgery. It does however seem logical, if the patient has symptomatic HCM with OTO and needs mitral valve surgery for IE, to perform septal myectomy at the same operation. It is a more contentious issue, if the patient was genuinely asymptomatic prior to the development of IE. Each case must of course be considered on an individual basis and with knowledge of the availability of local expertise.

### CONCLUSION

Infective endocarditis in HCM is an uncommon disease, but one which follows certain patterns. It is seen predominantly

in patients with OTO and left atrial enlargement. In view of the abnormal motion of the anterior mitral valve leaflet, it is the septal surface of this valve leaflet, which commonly holds vegetations. Depending on the clinical features, successful treatment can be accomplished with medical or surgical therapy. Depending on local familiarity and expertise, surgical therapy may include septal myectomy.

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