

Hypertrophic Cardiomyopathy: Experience in Pakistan

ISMAIL S. AHMED*, AZHAR M.A. FARUQUI** AND SHAUKET ALI SYED***

Hypertrophic Cardiomyopathy (HCM) is a disease or more likely a group of diseases characterized by hypertrophy of the ventricular musculature in the absence of any known stimulus for such hypertrophy. In the vast majority of cases, the hypertrophy is asymmetric i.e. the interventricular septum is thickened to a much

greater extent than the left ventricular free wall. In a small number of cases there is, however, concentric hypertrophy affecting the septum and the left ventricular free wall equally. In addition, there exists dynamic obstruction within the left ventricular outflow tract caused by a systolic partial re-opening of the anterior mitral leaflet into the left ventricular outflow already narrowed by the bulging septum (Fig. 1). While it affects mainly the left ventricle, in many cases there is hypertrophy and obstruction of the right ventricular outflow tract also.

HCM as an entity was recognized only 23 years ago (1). However, descriptions of this disease can be found in the pathological literature at the start of this century (2). In the over two decades since its recognition, there has been an explosion of papers regarding the various aspects of this disease.

First report on the existence of this disease in Pakistan was by one of us in 1978 (3). As the features of this disease entity as it exists in Pakistan have not been published in detail, we describe here our experience with the cases diagnosed at the National Institute of Cardiovascular Diseases (Pakistan), Karachi.

Material and Methods:

Between Jan'79 and July'80, a total of 460 Echoes were performed, out of which 13 patients were diagnosed to have HCM. Clinical history and a complete physical was obtained in all patients. ECG and Chest X-Ray P.A. View

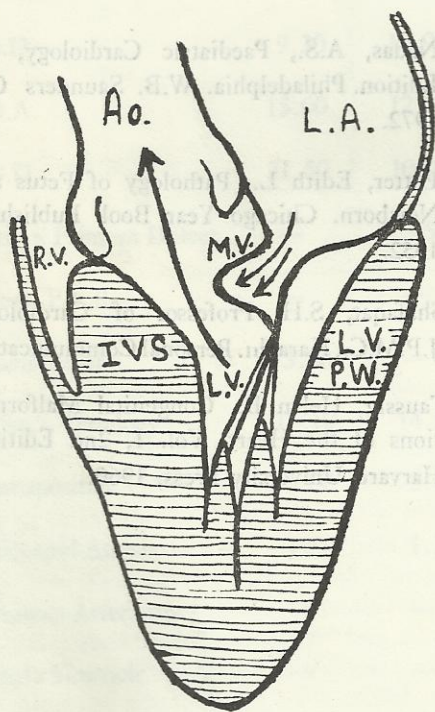


Fig. 1

*R.M.O., N.I.C.V.D. Karachi.

**Consultant, N.I.C.V.D., Karachi.

***Director, N.I.C.V.D., Karachi.

were done routinely. Echo was done using S&K EKOLINE A system with a 2.25 MHz transducer focused at 10 cm. Patients were examined in supine and left lateral positions. Measurements were made in the standard manner. HCM was diagnosed, when one of the following were present:

1. Asymmetric Septal Hypertrophy (ASH) i.e. Septal/Free Wall (Ratio ≥ 1.3).
2. Concentric L.V. Hypertrophy without any known etiology present.

Results:

Total No. of patients: 13.

Age 14-58 years (mean 36).

Sex: Males 11, Females 2.

Clinical Presentation:

Presenting Symptoms:

Chest Pain	6 Pts. (46%).
DOE	5 " (38%).
Palpitation	3 " (23%).
Syncopal Spells	1 " (7%).
N.B. Some Patients had multiple Symptoms.	

Cardiovascular Findings:

Bisferiens Pulse	3 Pts. (23%).
Systolic Murmur	9 " (69%).
Basal Crepitations	2 " (14%).

E.C.G. Changes:

Short PQ	1 Pts. (7%).
Abnormal Q Waves of Pseudo-infarction	6 " (46%).
L.V.H Pattern with increased voltage	7 " (60%).
R.V.H with tall R in VI	1 " (7%).
ST-T Changes	10 " (76%).

Note: No patient had normal E.C.G.

X-Ray Chest P.A. View:

CT Ratio = 0.5	6 Pts. (46%).
CT Ratio > 0.5 (Cardiomegaly)	4 Pts. (30%).
CT Ratio < 0.5	3 Pts. (23%).
Evidence of pulmonary venous congestion	2 Pts. (14%).

ECHO Features (Fig. 2):

ASH (IVS/LVPW ≥ 1.3)	12 Pts. (93%).
Concentric Left Ventricular Hypertrophy	1 " (7%).
SAM	10 " (76%).
Early Systolic Closure of the Aortic Valve	10 " (76%).
Flutter on the Aortic Valve Leaflets	11 " (83%).
Anteriorly Displaced Mitral Valve	12 " (93%).
(i.e. narrow L.V. Outflow)	

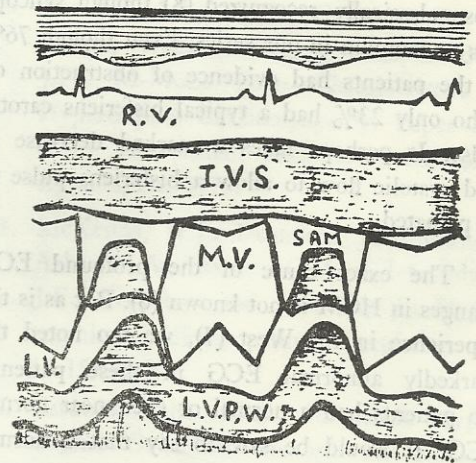


Fig. 2

Discussion:

It may seem odd that despite the tremendous interest HCM generated in the Cardiology circles all over the world, the disease was not described from Pakistan until 1978(3). The reason for this may have been partly a lack of awareness about this disease entity locally. However, the main reason perhaps was that the major diagnostic modality needed to firmly diagnose it i.e. Echocardiography was not introduced in Pakistan until 1978. Now that there is an increased sensitiveness towards diagnosing this condition and Echo facilities are available, there is atleast one new case of HCM diagnosed almost every month at the N.I.C.V.D., Karachi.

There seems a great preponderance of males in our series. While in general there is a male dominance in most reports from the West (4, 5), our male preponderance may be an artifact due to many possible reasons but none the less would suggest the same trend.

The presenting symptoms seen locally are those classically recognized (8) though syncope was uncommon in our series. Even though 76% of the patients had evidence of obstruction on Echo only 23% had a typical bisferiens carotid pulse. It perhaps takes a marked decrease in mid systolic flow to allow a bisferiens pulse to be palpated.

The exact cause of the profound ECG changes in HCM is not known (6). But as is the experience in the West (7), we too noted the markedly abnormal ECG in these patients. No patients had a normal or even near normal ECG. It would be safe to say that a normal ECG is a strong point against making the diagnosis of HCM.

The Chest X-Ray is of very little help diagnostically. It is however, a useful means of assessing pulmonary capillary pressure as in any other cardiac state. When cardiomegaly does occur, it is usually due to the dilatation of the right heart rather than the thick and stiff left ventricle which does not dilate until very late in the course of the disease (8).

While the presence of HCM was suspected in the majority of our cases, Echo remains the main means of confirmation. Presently, Cardiac Catheterization and angiography is almost never needed for diagnosis and are done either for research purposes or when surgery is contemplated in a patient who has failed to respond to medical therapy. Only one of our patients had cardiac cath. and angiography and that too as the diagnosis of valvular heart disease had ben made and Echo was at that time not locally available.

The main diagnostic feature in Echocardiography is the presence of asymmetric septal hypertrophy. Recently, more and more atypical cases of concentric hypertrophy are being recognized with other features of the disease much the same as in ASH (9). All cases showing systolic anterior motion of the mitral valve (SAM) also showed early sytolic closure of the aortic valve. They also exhibited flutter on the open aortic leaflets representing the early systolic obstruction to outflow and poor forward flow respectively. As such both SAM and mid-systolic closure of the aortic valve serve as accurate markers for following the severity of obstruction and response to therapy (10).

The long term natural history of HCM has been well documented in the West (11). Our local experience is must too short for meaningful analysis. In general, the response to pro-

propranolol orally has been very gratifying. All our patients are alive and improved. In one case, no symptomatic improvement occurred on propranolol. The dose of propranolol could not be increased beyond 120 mg daily due to concomitant presence of sick sinus syndrome and occurrence of severe sinus bradycardia. This patient was started on oral verapamil and has responded very well to it. No problem with sinus bradycardia has so far been noted on verapamil (12).

Summary:

Hypertrophic Cardiomyopathy occurs in Pakistan and is not very rare. Our experience with 13 cases is presented. In general, the features of the disease as seen locally are not much different from those seen elsewhere. Long term follow-up is needed to establish the natural history of the disease locally. Treatment with propranolol has had symptomatic improvement in all cases except one case which has responded to therapy with verapamil.

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