

**M E D I C A L   G R A N D   R O U N D S**

**UNIVERSITY OF TEXAS  
SOUTHWESTERN MEDICAL SCHOOL**

**HYPERTROPHIC CARDIOMYOPATHY (IHSS)**

**APRIL 14, 1988**

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HYPERTROPHIC CARDIOMYOPATHY

1. Introduction
2. Pathology
3. Clinical Symptoms
4. Physical Examination
5. Electrocardiogram
6. Chest Film
7. Echocardiography
8. Hemodynamic Studies
9. Natural History
10. Management
11. Yale Experience 1970-1988

The Dynamics of Left Ventricular  
Outflow Tract Obstruction

**Augmented Obstruction**

+Inotropy

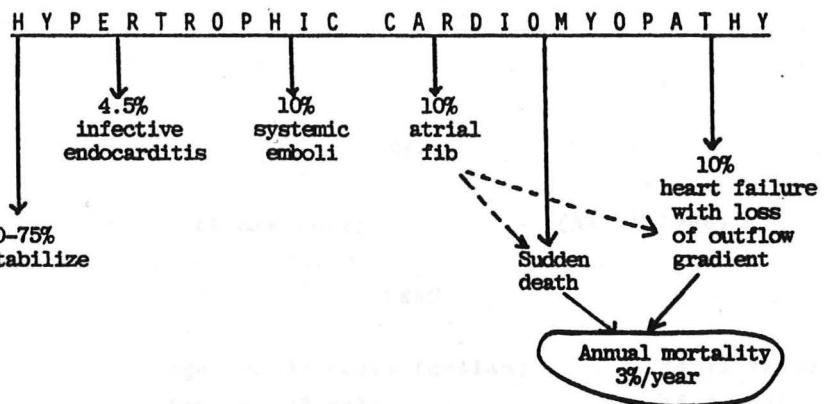
+LV Volume

+Aortic Diastolic  
Pressure

The Dynamics of Left Ventricular  
Outflow Tract Obstruction

Decreased Obstruction

- + Inotropy
- + LV Volume
- + Aortic Diastolic Pressure



HCM

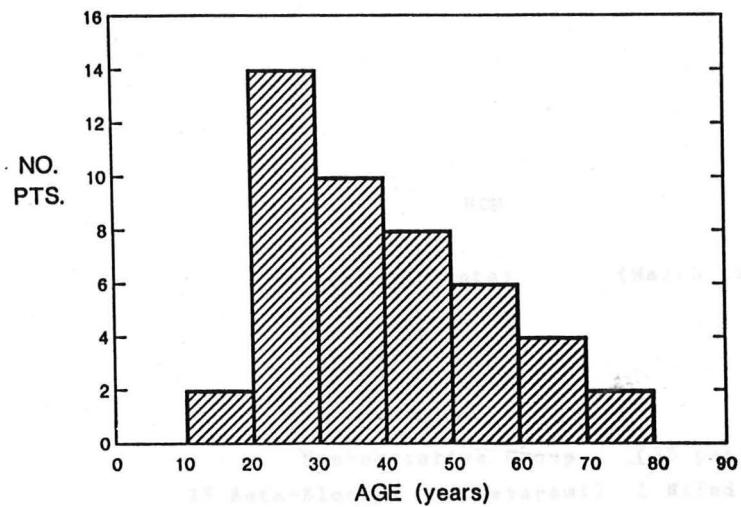
(46 patients)

(March 1988)

YNHH

Age - 37 years (median)	14-71 range
Sex - 32 male	14 female
Fam/Non Fam 14/32	
Followup 11 years (average)	1-18 range

**HYPERTROPHIC CARDIOMYOPATHY - YNHH**  
**March 1988**



HCM

(46 patients)

(March 1988)

YNHH

**Medications**

**Non-operative Group (35 patients)**

**25 Beta-Blocker 4 Verapamil 1 Nifedipine - 5 no meds**

**HCM**

**(46 patients) (March 1988)**

<b>Operation/no operation</b>	<b>11/35</b>
<b>Followup operation</b>	<b>11 years (average)</b>
	<b>1 - 21 range</b>

HCM

(46 patients)

(March 1988)

YNHH

Complications:

Subacute bacterial endocarditis	3
Hemolytic anemia	1
Pacemaker (post-op)	1
LBBB (post-op)	6

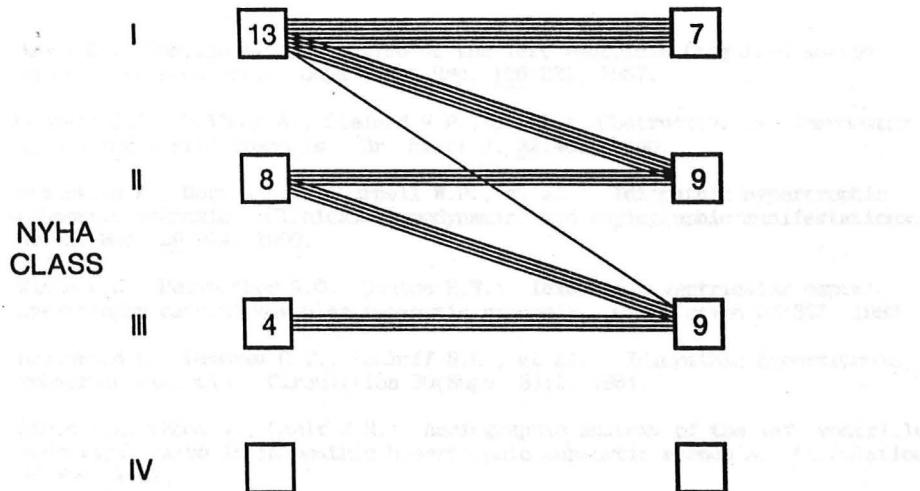
Alive/Dead\*

43/3

\*2 Sudden death

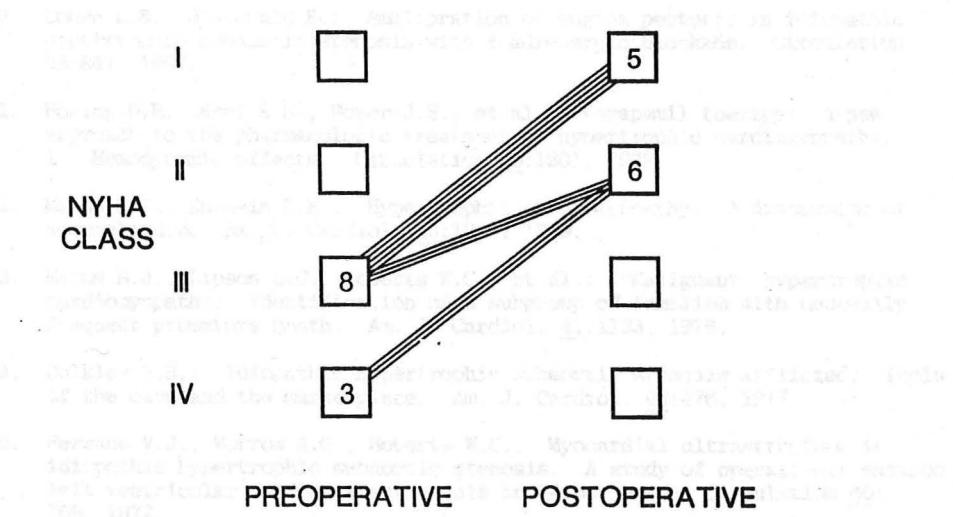
1 Pancreatic carcinoma

**HYPERTROPHIC CARDIOMYOPATHY - YNHH  
MEDICAL FOLLOW-UP (at least 3 years)  
March 1988**



# HYPERTROPHIC CARDIOMYOPATHY - YNHH

March 1988



REFERENCES

1. Brock R.: Functional obstruction of the left ventricle (acquired aortic subvalvular stenosis). *Guys Hosp. Rep.* 106:221, 1957.
2. Goodwin J.F., Hollman A., Clelard W.P., et al.: Obstructive cardiomyopathy simulating aortic stenosis. *Br. Heart J.* 22:403, 1960.
3. Braunwald E., Morrow A.G., Cornell W.P., et al.: Idiopathic hypertrophic subaortic stenosis. Clinical, hemodynamic, and angiographic manifestations. *Am. J. Med.* 29:924, 1960.
4. Wigle E.D., Heimbecker R.O., Gunton R.W.: Idiopathic ventricular septal hypertrophy causing muscular subaortic stenosis. *Circulation* 26:325, 1962.
5. Braunwald E., Lambrew C.T., Rockoff S.D., et al.: Idiopathic hypertrophic subaortic stenosis. *Circulation* 30(Supp. 3):1, 1964.
6. Simon A.L., Ross J., Gault J.H.: Angiographic anatomy of the left ventricle and mitral valve in idiopathic hypertrophic subaortic stenosis. *Circulation* 36:852, 1967.
7. Goodwin J.F., Oakley C.M.: The cardiomyopathies. *Br. Heart J.* 34:545, 1972.
8. Henry W.L., Clark C.E., Epstein S.E.: Asymmetric septal hypertrophy. Echocardiographic identification of the pathognomonic anatomic abnormality of IHSS. *Circulation* 42:225, 1973.
9. Clark C.E., Henry W.L., Epstein S.E.: Familial prevalence and genetic transmission of idiopathic hypertrophic subaortic stenosis. *N. Engl. J. Med.* 289:709, 1973.
10. Cohen L.S., Braunwald E.: Amelioration of angina pectoris in idiopathic hypertrophic subaortic stenosis with  $\beta$ -adrenergic blockade. *Circulation* 35:847, 1967.
11. Rosing D.R., Kent K.M., Borer J.S., et al.: Verapamil therapy: A new approach to the pharmacologic treatment of hypertrophic cardiomyopathy. I. Hemodynamic effects. *Circulation* 60:1201, 1979.
12. Maron B.J., Epstein S.E.: Hypertrophic cardiomyopathy: A discussion of nomenclature. *Am. J. Cardiol.* 43:1242, 1979.
13. Maron B.J., Lipson L.C., Roberts W.C., et al.: "Malignant" hypertrophic cardiomyopathy: Identification of a subgroup of families with unusually frequent premature death. *Am. J. Cardiol.* 41:1133, 1978.
14. Bulkley B.H.: Idiopathic hypertrophic subaortic stenosis afflicted: Idols of the cave and the marketplace. *Am. J. Cardiol.* 40:476, 1977.
15. Ferrans V.J., Morrow A.G., Roberts W.C.: Myocardial ultrastructure in idiopathic hypertrophic subaortic stenosis. A study of operatively excised left ventricular outflow tract muscle in 14 patients. *Circulation* 40:769, 1972.

16. Maron B.J., Roberts W.C.: Quantitative analysis of cardiac muscle cell disorganization in the ventricular septum of patients with hypertrophic cardiomyopathy. *Circulation* 59:689, 1979.
17. Frank S., Braunwald E.: Idiopathic hypertrophic subaortic stenosis. Clinical analysis of 126 patients with emphasis on the natural history. *Circulation* 37:759, 1966.
18. Wigle D.E., Adelman A.G., Auger P., et al.: Mitral regurgitation in muscular subaortic stenosis. *Am. J. Cardiol.* 24:698, 1969.
19. Shabetai R., Davidson S.: Asymmetrical hypertrophic cardiomyopathy simulating mitral stenosis. *Circulation* 45:37, 1972.
20. Savage D.D., Seides S.F., Clark C.E., et al.: Electrocardiographic findings in patients with obstructive and nonobstructive hypertrophic cardiomyopathy. *Circulation* 58:402, 1978.
21. Savage D.D., Slides S.F., Maron B.J., et al.: Prevalance of arrhythmias during 24-hour electrocardiographic monitoring and exercise testing in patients with obstructive and nonobstructive hypertrophic cardiomyopathy. *Circulation* 59:866, 1979.
22. McKenna W.J., Chetty S., Oakley C.M., Oakley C.M., et al.: Arrhythmia in hypertrophic cardiomyopathy: Exercise and 48-hour ambulatory electrocardiographic assessment with and without  $\beta$ -adrenergic blocking therapy. *Am. J. Cardiol.* 45:1, 1980.
23. Simon A.L.: Angiographic appearance of idiopathic hypertrophic subaortic stenosis. *Circulation* 46:614, 1972.
24. Brockenbrough E.C., Braunwald E., Morrow A.G.: A hemodynamic technic for the detection of hypertrophic subaortic stenosis. *Circulation* 23:189, 1961.
25. Braunwald E., Ebert P.A.: Hemodynamic alterations in idiopathic hypertrophic subaortic stenosis induced by sympathomimetic drugs. *Am. J. Cardiol.* 10:489, 1962.
26. Mason D.T., Braunwald E., Ross J.: Effects of changes in body position on the severity of obstruction to left ventricular outflow in idiopathic hypertrophic subaortic stenosis. *Circulation* 33:374, 1966.
27. Braunwald E., Oldham H.N., Ross, J., et al.: The circulatory response of patients with idopathic hypertrophic subaortic stenosis to nitroglycerin and to the Valsalva maneuver. *Circulation* 29:422, 1964.
28. Criley M.J., Lewis K.B., White R.I., et al.: Pressure gradients without obstruction. A new concept of "hypertrophic subaortic stenosis." *Circulation* 32:881, 1965.
29. Ross J., Braunwald E., Gault J.H., et al.: The mechanism of the intraventricular pressure gradient in idiopathic hypertrophic subaortic stenosis. *Circulation* 34:558, 1966.
30. Stewart S., Mason D.T., Braunwald E.: Impaired rate of left ventricular filling in idiopathic hypertrophic subaortic stenosis and ventricular aortic stenosis. *Circulation* 37:8, 1968.

31. Henry W.L., Clark C.E., Glancy D.L., et al.: Echocardiographic measurement of the left ventricular outflow gradient in idiopathic hypertrophic subaortic stenosis. *N. Engl. J. Med.* 288:989, 1973.
32. Maron B.J., Epstein S.E.: Hypertrophic cardiomyopathy. Recent observations regarding the specificity of three hallmarks of the disease: Asymmetrical septal hypertrophy, septal disorganization and systolic anterior motion of the anterior mitral leaflet. *Am. J. Cardiol.* 45:141, 1980.
33. Martin R.P., Rakowski H., French J., et al.: Idiopathic hypertrophic subaortic stenosis viewed by wide-angle, phased-array echocardiography. *Circulation* 59:1206, 1979.
34. Maron B.J., Gottdiener J.S., Epstein S.E.: Patterns and significance of the distribution of left ventricular hypertrophy in hypertrophic cardiomyopathy. A wide-angle two dimensional echocardiographic study of 125 patients. *Am. J. Cardiol.* 48:418, 1981.
35. Rubin K.A., Morrison J., Paonick M.B., et al.: Idiopathic hypertrophic subaortic stenosis: Evaluation of anginal symptoms with thallium-201 myocardial imaging. *Am. J. Cardiol.* 44:1040, 1979.
36. Pohost G.M., Vignola P.A., McKusick K.E., et al.: Hypertrophic cardiomyopathy. Evaluation by gated cardiac blood pool scanning. *Circulation* 55:92, 1977.
37. Borer J.S., Bacharach S.L., Green M.V., et al.: Effect of septal myotomy and myectomy on left ventricular systolic function at rest and during exercise in patients with IHSS. *Circulation* 60:I-82, 1979.
38. Bonow R.O., Rosing O.R., Bacharach S.L., et al.: Effect of verapamil on left ventricular systolic function and diastolic filling in patients with hypertrophic cardiomyopathy: Assessment with radionuclide cineangiography. *Circulation* 64:787, 1981.
39. Hardarson T., De La Calzada C.S., Curiel R., et al.: Prognosis and mortality of hypertrophic obstructive cardiomyopathy. *Lancet* 2:1462, 1973.
40. Shah P.M., Adelman A.G., Wigle E.D., et al.: The natural (an unnatural) history of hypertrophic obstructive cardiomyopathy. *Circ. Res.* 34, 35(Supp. II):179, 1974.
41. Wang K., Gobel F.L., Gleason D.F.: Bacterial endocarditis in idiopathic hypertrophic subaortic stenosis. *Am. Heart J.* 89:359, 1975.
42. McKenna W., Deanfield J., Faruqui A., et al.: Prognosis in hypertrophic cardiomyopathy: Role of age and clinical, electrocardiographic and hemodynamic features. *Am. J. Cardiol.* 47:532, 1981.
43. Maron B.J., Roberts W.C., Edwards J.E., et al.: Sudden death in patients with hypertrophic cardiomyopathy: Characterization of 26 patients without functional limitation. *Am. J. Cardiol.* 41:803, 1978.

44. Adelman A.G., Shah P.M., Gramiak R., et al.: Long-term propranolol therapy in muscular subaortic stenosis. Br. Heart J. 32:804, 1970.
45. Stenson R.E., Flamm M.D., Harrison D.C., et al.: Hypertrophic subaortic stenosis. Clinical and hemodynamic effects of long-term propranolol therapy. Am. J. Cardiol. 31:763, 1973.
46. Rosing D.R., Kent K.M., Maron B.J., et al.: Verapamil therapy: A new approach to the pharmacologic treatment of hypertrophic cardiomyopathy. II. Effects on exercise capacity and symptomatic status. Circulation 60:1208, 1979.
47. Kaltenbach M., Hopf R., Kober G., et al.: Treatment of hypertrophic obstructive cardiomyopathy with verapamil. Br. Heart J. 42:35, 1979.
48. Morrow A.B., Brockenbrough E.C.: Surgical treatment of idiopathic hypertrophic subaortic stenosis. Technic and hemodynamic results of subaortic ventriculotomy. Ann. Surg. 154:181, 1961.
49. Wigle E.D., Chrysohou A., Bigelow W.G.: Results of ventriculotomy in muscular subaortic stenosis. Am. J. Cardiol. 11:572, 1963.
50. Morrow A.G.: Hypertrophic subaortic stenosis: Operative methods utilized to relieve left ventricular outflow obstruction. J. Thorac. Cardiovasc. Surg. 76:423, 1978.
51. Maron B.J., Merrill W.H., Frier P.A., et al.: Long-term clinical course and symptomatic status of patients after operation for hypertrophic subaortic stenosis. Circulation 57:1205, 1978.
52. Redwood D.R., Goldstein R.E., Hirshfeld J., et al.: Exercise performance after septal myotomy and myectomy in patients with obstructive hypertrophic cardiomyopathy. Am. J. Cardiol. 44:215, 1979.
53. Maron B.J., Savage, D.D., Wolfson, J.K., et al.: Prognostic Significance of 24 Hour Ambulatory Electrocardiographic Monitoring in Patients with Hypertrophic Cardiomyopathy: A Prospective Study. Am. J. Cardiol. 48:252, 1981.
54. Gilbert B.W., Pollick, C., Adelman, A.G., et al.: Hypertrophic Cardiomyopathy: Subclassification by M Mode Echocardiography. Am. J. Cardiol. 45:861, 1980.
55. Gardin J.M., Talano, J.V., Stephanides, L., et al.: Systolic Anterior Motion in the Absence of Asymmetric Septal Hypertrophy. Circulation 63:181, 1981.
56. Pollick C., Morgan, C.D., Gilbert, B.W., et al.: Muscular Subaortic Stenosis: The Temporal Relationship between Systolic Anterior Motion of the Anterior Mitral Leaflet and the Pressure Gradient. Circulation 66:1087, 1982.

57. McKenna W.J., Borggrefe, M., England, D., et al.: The Natural History of Left Ventricular Hypertrophy in Hypertrophic Cardiomyopathy: An Electrocardiographic Study. *Circulation* 66:1233, 1982.
58. Lorell B.H., Paulus, W.J., Grossman, W., et al.: Modification of Abnormal Left Ventricular Diastolic Properties by Nifedipine in Patients with Hypertrophic Cardiomyopathy. *Circulation* 65:499, 1982.
59. Murgo J.P., Alter, B.R., Dorothy, J.F., et al.: Dynamics of Left Ventricular Ejection in Obstructive and Nonobstructive Hypertrophic Cardiomyopathy. *J. Clin. Invest.* 66:1369, 1980.
60. Shah P, Taylor RD, Wong M: Abnormal mitral valve coaptation in hypertrophic obstructive cardiomyopathy: Proposed role in systolic anterior motion of mitral valve. *Am J Cardiol* 48:258-262, 1981.
61. Spirito P, Maron B, Rosing D: Morphologic determinants of hemodynamic state after ventricular septal myotomy-myectomy in patients with obstructive hypertrophic cardiomyopathy: M-mode and two-dimensional echocardiographic assessment. *Circulation* 70:984-995, 1984.
62. Pollici C, Williams WG, Rakowski H. et al.: Post-ventriculomyectomy eccentric SAM: Further evidence for the Venturi mechanism. *J Amer Coll Cardiol* 3:492, 1984 (abstr).
63. Goodwin JF: The frontiers of cardiomyopathy. *Br Heart J* 48:1-18, 1982.
64. Cannon RO, Rosing DR, Maron BJ, et al: Myocardial ischemia in hypertrophic cardiomyopathy: Contribution of inadequate vasodilator reserve and elevated left ventricular filling pressures. *Circulation* 71:234-243, 1985.
65. Bertrand ME, Tilman PY, Lablanche JM, et al: Apical hypertrophic cardiomyopathy: Clinical and metabolic studies. *Eur Heart J* 4:127-133, 1983 (Suppl F).
66. Lorell BH, Paulus WJ, Grossman W: Modification of abnormal left ventricular diastolic properties by nifedipine in patients with hypertrophic cardiomyopathy. *Circulation* 65:499-507, 1982.
67. Lorell BH, Paulus WJ, Grossman W, et al: Improved diastolic function and systolic performance in hypertrophic cardiomyopathy after nifedipine. *New Engl J Med* 303:801-803, 1980.

68. Maron BJ, Wolfson JK, Ciro E, et al: Relation of electrocardiographic abnormalities and patterns of left ventricular hypertrophy identified by 2-dimensional echocardiography in patients with hypertrophic cardiomyopathy. Am J Cardiol 51:189-194, 1983.
69. Maron BJ, Bonow RO, Seshagiri TN, et al: Hypertrophic cardiomyopathy with ventricular septal hypertrophy localized to the apical region of the left ventricle (apical hypertrophic cardiomyopathy). Am J Cardiol 49:1838-1848, 1982.
70. Suwa M, Hirota Y, Kawamura K: Improvement in left ventricular diastolic function during intravenous and oral diltiazem therapy in patients with hypertrophic cardiomyopathy: An echocardiographic study. Am J Cardiol 54:1047-1053, 1984.
71. Redwood DR, Goldstein RE, Hirshfeld J, et al: Exercise performance after septal myotomy and myectomy in patients with obstructive hypertrophic cardiomyopathy. Am J Cardio 44:215-220, 1979.
72. Beahrs MM, Tajik AJ, Seward JB, et al: Hypertrophic obstructive cardiomyopathy: 10-21 year follow-up after partial septal myectomy. Am J Cardiol 51:1160-1166, 1983.
73. Isner JM, Clarke RH, Pandian NG, et al: Laser myoplasty for hypertrophic cardiomyopathy. In vitro experience in human post-mortem hearts and in vivo experience in a canine model (transarterial) and human patients (intraoperative). Am J Cardiol 53:1620-1625, 1984.
74. McKenna W, Deanfield J, Faruqui A, et al: Prognosis in hypertrophic cardiomyopathy: Role of age and clinical electrocardiographic and hemodynamic features. Am J Cardiol 47:532-538, 1981.
75. McKenna W, Harris L, Deanfield J: Syncope in hypertrophic cardiomyopathy. Br Heart J 47:177-179, 1982.
76. McKenna WJ, Chetty S, Oakley CM, et al: Arrhythmia in hypertrophic cardiomyopathy: Exercise and 48 hour ambulatory electrocardiographic assessment with and without beta-adrenergic blocking therapy. Am J Cardiol 45:1-5, 1980.
77. McKenna W, Harris L, Rowland E, et al: Amiodarone for long-term management of patients with hypertrophic cardiomyopathy. Am J Cardiol 54:802-810, 1984.
78. Ciro E, Maron BJ, Bonow RO, et al: Relation between marked changes in left ventricular outflow tract gradient and disease progression in hypertrophic cardiomyopathy. Am J Cardiol 53:1103-1109, 1984.